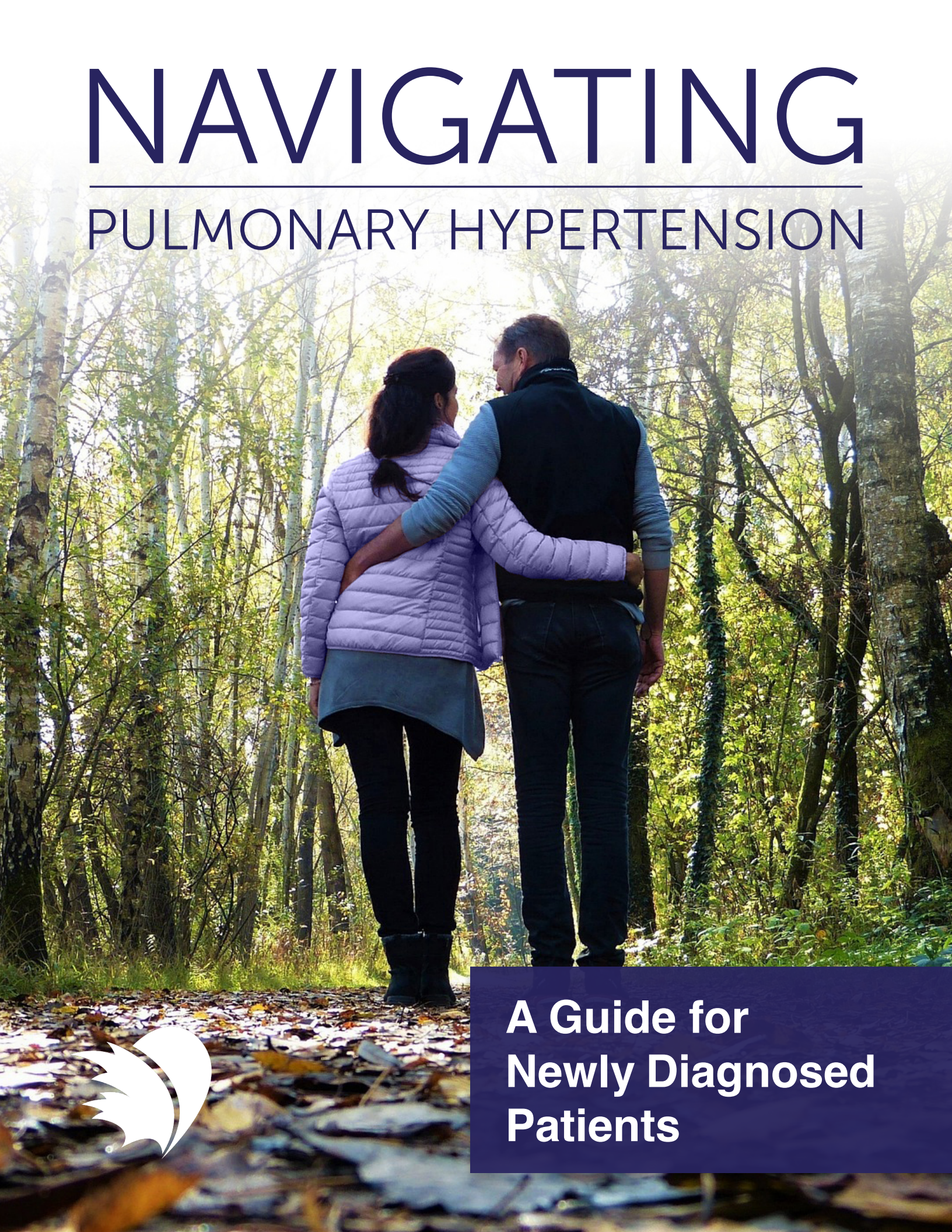
NAVIGATING

PULMONARY HYPERTENSION



A Guide for Newly Diagnosed Patients

Version

Version 2, Published January 2025

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Welcome to the Pulmonary Hypertension Community

You and your loved ones probably have a lot of questions and emotions after receiving a pulmonary hypertension (PH) diagnosis. This resource aims to answer those questions and help you learn more about PH.

The Pulmonary Hypertension Association of Canada (PHA Canada) provides this publication to help you find education, support, and hope during the weeks, months, and years after diagnosis. Although this guide won't address every question you encounter in your journey, it provides an overview of diagnosis, treatment, symptoms, and quality of life issues. You'll find resources and questions to ask your doctor and other information about PH. You'll also learn about ways to tap into a network of support, become involved in research, and address some of the many issues that come up in day-to-day living with a chronic disease.

We don't expect you to read the entire publication from beginning to end at once. Instead, take your time to browse the contents and select the sections that are meaningful to you on any given day. Take your time, be gentle with yourself and others, and contact PHA Canada or your PH team anytime you have a question or feel worried, unsure, or overwhelmed.

Your PH journey is unique to you. If this guide seems overwhelming, set it aside and return to it another time. When you're ready to pick it up again, use the notes section to write down additional questions you have for your care team and support network.

PHA Canada is here to help you. The association was founded in 2008 by patients, caregivers, and healthcare professionals to work together to better the lives of all Canadians affected by pulmonary hypertension and represent a united national PH community. Whether you have PH or help care for someone with PH, Navigating Pulmonary Hypertension: A Guide for Newly Diagnosed Patients will help you navigate this new journey.

If you or a family member/caregiver need to ask questions or get support, you can reach us at **info@phacanada.ca** or 1-877-774-2226.

PULMONARY HYPERTENSION ASSOCIATION OF CANADA

L'ASSOCIATION D'HYPERTENSION PULMONAIRE DU CANADA

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What is Pulmonary Hypertension?

Pulmonary hypertension (PH) is a complex, often misunderstood disease. PH simply means high blood pressure in the lungs. With PH, blood vessels in the lungs become stiff and narrow, impeding blood flow. As a result, the right side of the heart must work harder to pump blood.

Normal pressure in the arteries of the lungs (called "pulmonary artery pressure") ranges from 8 to 20 mmHg at rest. People with PH generally have an average resting pulmonary artery pressure above 20 mmHg.*

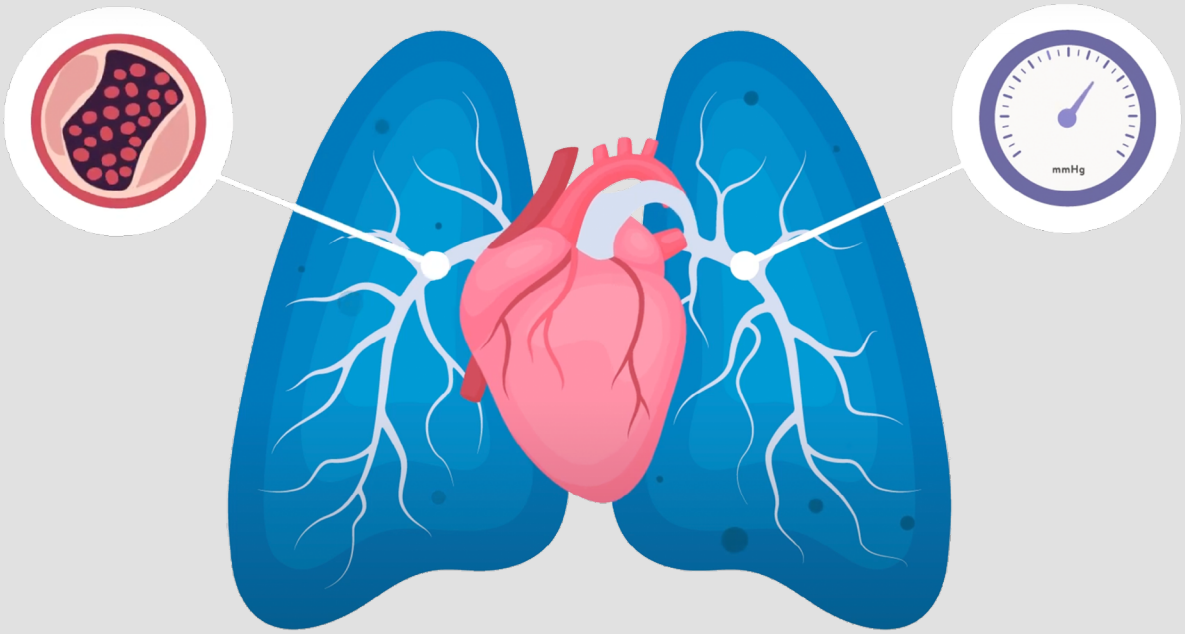
PH differs from "regular" – or systemic – hypertension, which most people refer to as high blood pressure. With systemic hypertension, pressure is higher than it should be in arteries throughout the body. With PH, pressure is higher in the lungs. In most cases, systemic hypertension and pulmonary hypertension are completely unrelated.

PH affects people of all ages and backgrounds. Some people are diagnosed with PH as infants or children, and others as teens, adults, or seniors.

There are five types (or "groups") of PH, each based on the underlying causes. Each group is different, as is each patient. It's important for newly diagnosed patients to find appropriate care as soon as possible after diagnosis to pinpoint the cause and develop a treatment plan.

People with PH can live long, meaningful lives with the right care and treatment. In fact, advances in care and treatment have improved the outlook for people living with this disease and, one day, might lead to a cure.

Keep reading, and be sure to watch our **Introduction to PH** video series at **phacanada.ca/videos**



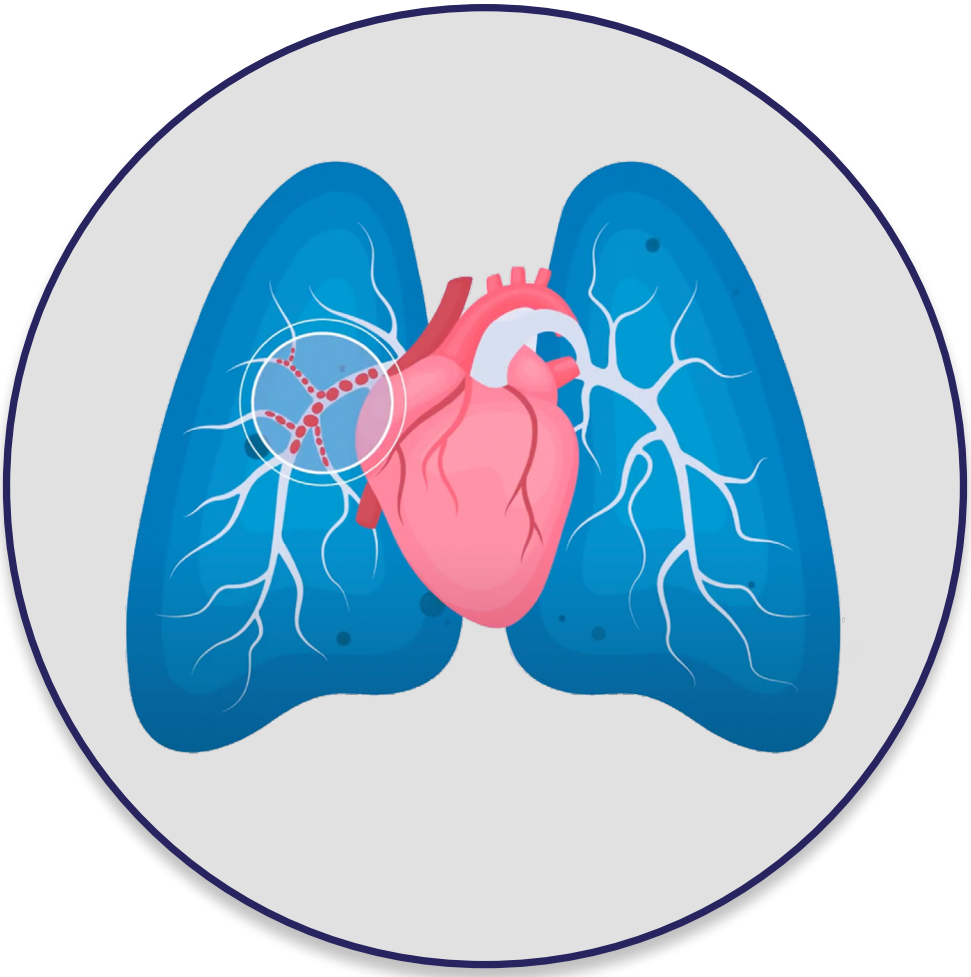
** Simonneau G, et al. Eur Respir J. 2019;53(1):1801913.*

Heart and Lungs: Understanding the Circulatory System

To better understand why PH can be so serious, it's helpful to understand how blood circulates between the heart and lungs. The heart and lungs work together to carry oxygen throughout the body. The heart is a muscle made up of two pumping chambers called ventricles – the right and left ventricles – that pump blood to different areas of the body. As blood returns from the rest of the body, it first goes into the right ventricle, which pumps it into the lungs.

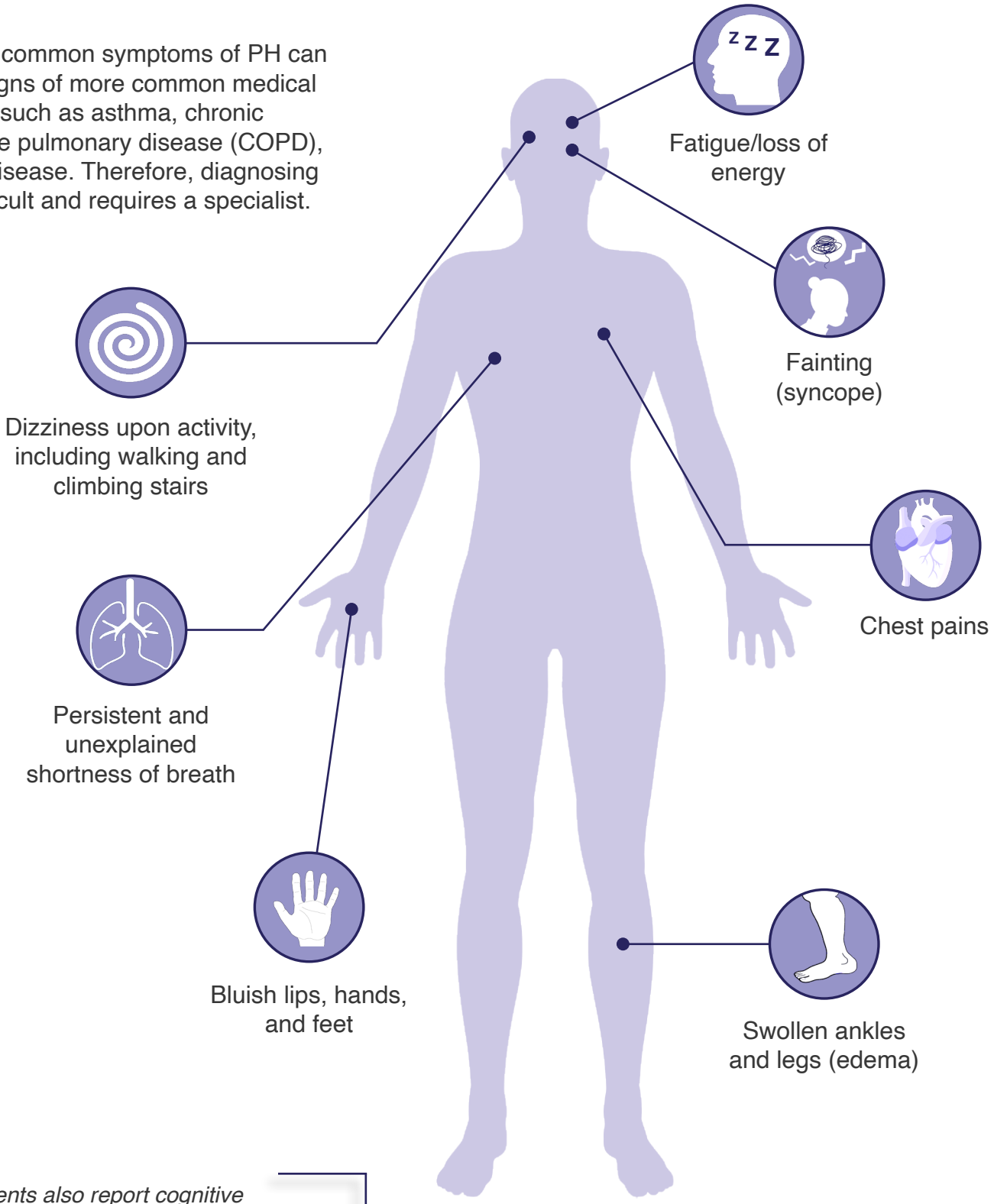
The lungs take carbon dioxide from the blood — which the body releases as you exhale — and replaces it with the oxygen you inhale. After the blood picks up oxygen, it is considered "oxygenated" and is ready to return to other areas of the body. The blood then travels from the lungs to the left side of the heart. The left ventricle pumps blood to the rest of the body, where it can deliver oxygen to your muscles, and the process starts over again with each heartbeat.

The left ventricle is more muscular because it must pump blood to the entire body. To send blood from your head to your toes and back again, a higher-pressure system is needed. The right side of the heart is smaller and less muscular than the left side because it pumps blood only through the lungs, normally a low-pressure system.



Common Symptoms

The most common symptoms of PH can also be signs of more common medical problems such as asthma, chronic obstructive pulmonary disease (COPD), or heart disease. Therefore, diagnosing PH is difficult and requires a specialist.



Some patients also report cognitive symptoms, such as minor memory issues and "brain fog." Those issues could be related to problems PH causes in the circulatory system, such as preventing oxygenated blood from freely flowing throughout the body, including the brain.*

**Brown LM, et al. CHEST. 2011;140(1):19-26.*

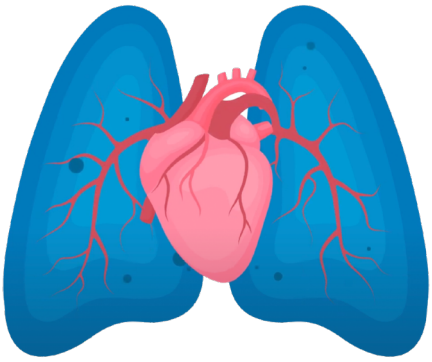
Types of Pulmonary Hypertension

Not all PH is the same. PH is a general term for high blood pressure in the lungs from any cause. There are five groups of PH based on different causes. Physicians review the classifications every five years at the World Symposium on Pulmonary Hypertension. This classification was originally developed by the World Health Organization, so sometimes you might see references to "WHO Groups." Each type of PH has different causes and treatments that help people with PH live a better life.

Group 1

Group 1 refers to **pulmonary arterial hypertension (PAH)**. It is caused when arteries in the lungs narrow, thicken, and become rigid. The right side of the heart must work harder to push blood to the lungs through those arteries. The extra stress can cause the heart to lose the ability to pump enough blood throughout the lungs to meet the needs of the rest of the body.

There are several types of PAH. Idiopathic PAH (IPAH) is the name given to the disease when no clear cause can be found. PAH can also develop in association with other medical conditions, including congenital heart disease, liver disease, HIV, and connective tissue diseases such as scleroderma and lupus. PAH can also be associated with previous or ongoing use of some drugs, including methamphetamine or certain diet pills.



Heritable PAH (HPAH) is a form of pulmonary hypertension that can be inherited and/or passed on. There is often a family history of PAH. Of the people who carry a gene associated with an increased risk of PH, not everyone will develop the disease. Genetic testing can help you find out whether you carry one of those genes. Before pursuing genetic testing, discuss the pros and cons with your PH specialist.

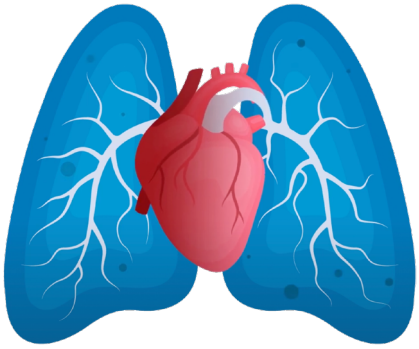
PVOD (pulmonary veno-occlusive disease) and PCH (pulmonary capillary hemangiomatosis) are very rare types of PAH that involve the small veins and capillaries (tiny vessels found between arteries and veins of the lungs) of the lungs. Diagnosis and treatment of PVOD and PCH can be uniquely challenging. Currently, lung transplantation is the best treatment available.

While several treatment options are available for PAH, there is no known cure.

Some people use the terms PH and PAH interchangeably, but they aren't the same. All PAH is PH, but not all PH is PAH.

Group 2

Group 2 PH is the most common form of PH in developed countries. It refers to **PH due to left-sided heart disease**. Diseases of the left side of the heart result in high pressures in the left side of the heart, which causes a back-up of blood (and pressure) in the lungs, resulting in PH. Common risk factors for Group 2 PH include heart valve disease, heart failure, as well as diabetes, systemic hypertension, obesity, and atrial fibrillation.

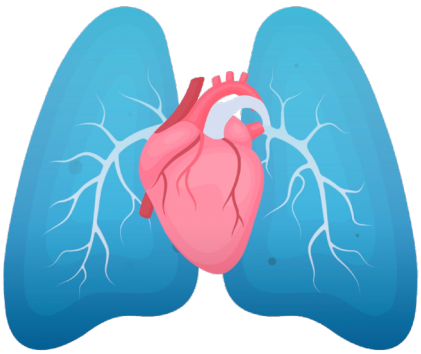


Improving left heart function can also improve Group 2 PH.

Group 3

Group 3 refers to **PH due to chronic lung disease and/or hypoxia** (low oxygen levels). Examples of common conditions that can cause this are:

- Obstructive lung disease, where the lung airways narrow and make it harder to exhale (e.g., COPD/emphysema).
- Restrictive lung disease, in which the lungs have a tough time expanding with inhalation (e.g., interstitial lung disease, pulmonary fibrosis).
- Other Group 3 causes include:
  - Severe forms of obstructive sleep apnea (e.g., obesity hypoventilation syndrome).
  - Living at a high altitude for a long time. In this situation, arteries in the lungs tighten, so blood goes only to areas of the lungs that receive the most air and oxygen. Over time, that tightening leads to high blood pressure throughout the lungs.

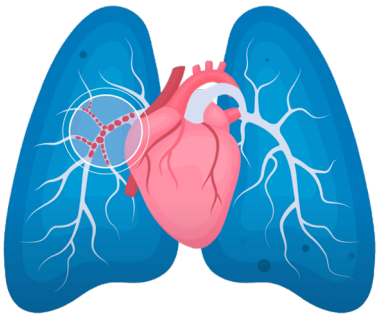


Group 3 PH can be improved by treating underlying causes.

Group 4

Group 4 PH refers to **PH associated with pulmonary artery obstructions. Chronic thromboembolic pulmonary hypertension (CTEPH)** is the most common disease in this category. This occurs when the body can't dissolve a blood clot ("pulmonary embolism" or "PE") in the lungs. Undissolved clots can cause scarring in the blood vessels of the lungs. The scarring impedes blood flow and makes the right side of the heart work harder than usual. Not all patients who have CTEPH will have a history of previous PE, however.

CTEPH differs from other forms of PH because it can potentially be cured. For that reason, people with PH should be screened for CTEPH with a test called a ventilation-perfusion scan (V/Q Scan).



Some CTEPH patients are eligible for pulmonary thromboendarterectomy (PTE) surgery, which removes blood clots and scar tissue that cause CTEPH. However, the surgery isn't suitable for all people with CTEPH.

If a doctor determines someone isn't a candidate for PTE surgery, the patient might be able to have a balloon pulmonary angioplasty (BPA) or medications to open the lung blood vessels. Others might need more than one treatment. Your PH specialist team, along with CTEPH experts, will work together to provide a tailored treatment plan.

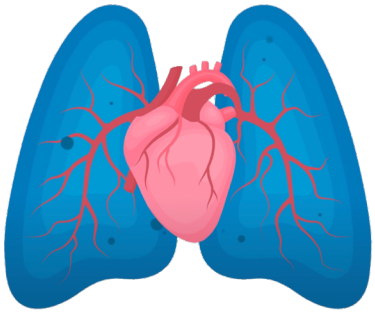
In some cases, PH remains after PTE surgery. Those patients also might be treated with BPA or medication.

All CTEPH patients should be evaluated at a specialized CTEPH center.

Learn more about CTEPH at **phacanada.ca/CTEPH**

Group 5

Group 5 refers to **PH that occurs with unclear and/or multifactorial mechanisms**. In those cases, PH is secondary to the associated conditions in ways that aren't well understood. The associated conditions include but aren't limited to sarcoidosis, sickle cell anemia, chronic hemolytic anemia, end-stage kidney disease, and some metabolic disorders.



No matter the type, PH is a serious disease. If untreated, PH can lead to right heart failure and death. Read more about the types of PH at **phacanada.ca/typesofph**

PH Risk Factors

A PH diagnosis can make people worry. You and your family may be wondering why this happened to you. What puts someone at risk of getting PH? People of all ages, sexes, and backgrounds can be diagnosed with PH. However, certain medical conditions and risk factors can make some people more likely to get PH than others.

- **Family History:** When two or more immediate family members (parent, sibling, child) have PAH, or a family member is known to have a PAH-causing gene.
- **Obesity:** Obesity is a risk factor for PH and can cause PH through several mechanisms.
- **Gender:** Some types of PAH are more common in women than in men.
- **Drugs and Toxins:** Certain drugs are known to cause pulmonary arterial hypertension (PAH).
- **Other diseases:** Many other medical conditions may also cause PH.

Learn more about risk factors and associated conditions for PH at **phacanada.ca/riskfactors**.

Sometimes we don't know why someone develops PH. What we do know is that you are not alone on this journey. There is an amazing, compassionate community standing behind you. You probably have more questions.

Find answers to some of the most common questions about pulmonary hypertension on our FAQ page at **phacanada.ca/FAQ**

PH Diagnosis

PH can be difficult to diagnose in routine medical exams because the most common symptoms of PH (breathlessness, fatigue, and dizziness) are also associated with many more common conditions.

When doctors suspect someone has PH, they will review the patient's medical and family history and perform physical exams and diagnostic tests.

PH is often late to be diagnosed. Medical awareness of PH is improving, but it remains much less common than other heart and lung diseases that can cause similar symptoms. The most important step in diagnosing PH is being aware that it is a possibility and undergoing the key screening test for PH: an echocardiogram.

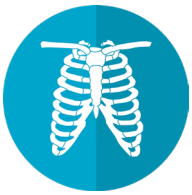
When doctors suspect someone has PH, they review the patient's medical and family history and perform physical exams and diagnostic tests. Before conducting diagnostic tests, your doctor will conduct a physical examination. The doctor will look for visible and/or enlarged veins along the sides of the neck, irregular heart sounds, and swelling in your abdomen, legs, or ankles (edema). The doctor then will perform a series of tests to identify your type of PH and what might have caused it; this will guide your treatment plan.

Diagnostic tests include blood tests, chest x-rays, electrocardiograms (EKG), echocardiograms, pulmonary function tests (PFTs), exercise tolerance tests (six-minute walk or cardiopulmonary exercise), and CT scans. But the only way to definitively diagnose PH is through right heart catheterization.

Once I finally dragged myself to the doctor the stars were aligned in my favour. From my family doctor to the PH clinic with dozens of tests in between I was diagnosed and started treatment in less than 4 months . The first place I was directed to for information was PHA Canada. I will be forever grateful for the soft landing that they afforded me. Their resources and guidance got me comfortable on my PH journey.

Marion Roth
Living with PAH since 2011
Milverton, ON

PH Detection Tests



Chest X-rays can reveal an enlarged right ventricle or pulmonary arteries. Chest X-rays can also show signs of emphysema or scarring (interstitial fibrosis) of the lungs.

An **Electrocardiogram (ECG)** checks the electrical impulses of the heart. Electrodes are attached to the patient's skin, and a recording of these impulses is made. However, an ECG alone is not enough to indicate a PH diagnosis. If your doctor performs an ECG, they will also perform additional procedures to identify PH.

An **Echocardiogram (or "echo")** is a procedure where electrodes are placed on the patient's skin, and an ultrasound of the heart is taken. This painless procedure is often used to make a preliminary diagnosis by estimating the pressures in the right heart and assessing how well the heart is functioning. Other heart conditions that produce symptoms similar to PH may be diagnosed with an echocardiogram. Additionally, an echocardiogram may be used to monitor a patient's condition and response to treatment over time.

PH Group Identification Tests

Pulmonary Function Tests measure how much air your lungs can hold, how much air moves in and out of them, and the lungs' ability to exchange oxygen. These tests may be performed to potentially identify the cause of PH.

Computerized Tomography (CT) Chest Scan is a non-invasive test that uses small amounts of radiation to create many precise pictures of the structures in the chest, including the heart and lungs. Sometimes, a dye (also known as "contrast") is injected into the arm to make the images of certain structures clearer. The test allows healthcare providers to see the lungs in far greater detail than with chest X-rays. Doctors can see the size of the lungs, larger blood vessels in the lungs, and lung tissue. The test looks for evidence of chronic lung diseases (Group 3 PH) or blood clots (Group 4 PH).

Nuclear Scan (Ventilation/Perfusion Scan or V/Q Scan) is a diagnostic tool which tests for blood clots in the lungs by producing a picture of air and blood flow to the lungs. A small dose of radioactive material is breathed in, and another small dose is injected via a blood vessel into the lungs. The doctor will review the images that are produced to evaluate the health of the lungs. This test is recommended to rule out CTEPH in all cases of PH.

Blood Tests check oxygen levels in the blood (i.e., with an arterial blood gas), observe liver and kidney function, and identify whether the patient has connective tissue disease (such as lupus or scleroderma), thyroid problems, or signs of infection or HIV. To help assess the function of the heart and to monitor response to treatment, a brain natriuretic peptide (BNP) test or an N-Terminal-pro brain natriuretic peptide (NT-proBNP) may be conducted. Some centers measure NT-proBNP while others measure BNP, but both measure the same thing – heart function.

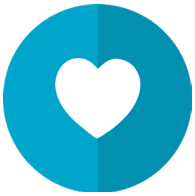
Sleep Studies are occasionally requested in patients at risk for obstructive sleep apnea or low oxygen levels overnight. These can be done at home with equipment to check for snoring and oxygen levels, or may require a complete polysomnogram, where a patient sleeps in a monitored sleep lab.

Exercise Capacity Test

A **Six-Minute Walk Test (6MWT)** measures how far a patient can walk in six minutes. It also measures perceived exertion, heart rate, and oxygen saturation (how much oxygen is in the blood). Doctors compare test results from each clinic visit as one way to measure whether PH is improving or worsening.

A **Cardiopulmonary Exercise Test (CPET)** is used to measure blood pressure and oxygen consumption when patients exercise (often on a stationary bike). The test identifies how well both heart and lungs respond to exercise.

PH Hemodynamics: Testing The Heart Muscle



During a **Right Heart Catheterization** (RHC or "right heart cath"), a doctor inserts a small tube into a vein in the neck, upper arm, or groin and guides it into the right atrium of the heart. The tube is then guided through the right ventricle into the pulmonary artery. The practitioner will then take several pressure readings and sometimes withdraw blood from the catheter tip.

The doctor will calculate how much blood the heart can pump in a minute (cardiac output) and take a reading called a "wedge" pressure. In this test, a balloon is inflated at the end of the catheter and wedged into a smaller section of one of the pulmonary arteries. The reading gives insight into the pressure on the left side of the heart and can indicate left heart disease that contributes to Group 2 PH. Patients with PAH (Group 1 PH) will have normal wedge pressure.

> The only way to definitively diagnose PAH is through right heart catheterization.

Many of the tests used to diagnose PH can also help identify a patient's "functional class". Your providers might require a right heart catheterization periodically or a six-minute walk test at every visit. Those repeated tests help them evaluate your health and – with your input – make the best decisions about your treatment and care. Some reasons doctors order these tests include changes to your health status, events like hospitalizations, or medication changes. If you ever wonder why a test is being run or what the results mean, ask your PH specialist.

> After doctors confirm a PH diagnosis and identify the type, they will use other tests to determine how best to treat a patient's PH, monitor how they respond to treatments, and obtain other information to forecast disease progression.

Learn more about the tests involved in diagnosing PH at **phacanada.ca/diagnosis**

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Treatment Options

Advancements in the past three decades have led to medical therapies for pulmonary arterial hypertension and CTEPH that target the pulmonary arteries (PH-targeted therapies). Medication can relieve symptoms, improve quality of life, and slow disease progression. Your treatment options depend on your type of pulmonary hypertension, your functional class (see page 24), other medications you take, and other conditions you have, among other considerations.

Many treatments target **pulmonary arterial hypertension (PAH, Group 1 PH)** and more are in the drug-development pipeline. Treatments include conventional medical therapies such as diuretics ("water pills") and oxygen therapy. Ordinary blood pressure pills called calcium channel blockers can be very effective for a small group of patients. Therapies for PAH include oral, inhaled, subcutaneously injected, subcutaneously infused (under the skin), and intravenously infused (into the vein) medications. See the next page for a list. Not all treatments available worldwide are accessible in Canada (e.g. inhaled treprostinil).

Group 2 pulmonary hypertension (due to left-sided heart disease) and Group 3 pulmonary hypertension (due to chronic lung disease and/or hypoxia) are managed by treating the underlying heart or lung condition. There is emerging research into PH-specific therapies for these conditions.

People with **CTEPH (Group 4 pulmonary hypertension)** can benefit from surgery to remove blood clots and scar tissue in the lungs. Patients who aren't candidates for surgery or whose pulmonary hypertension remains after surgery might benefit from balloon pulmonary angioplasty (BPA) or PH-specific medication.

Depending on the severity of your pulmonary hypertension, a lung or heart-lung transplant might also be an option.

Each patient is different. Talk to your pulmonary hypertension doctor about the best treatments for your specific diagnosis.

Pulmonary hypertension is a progressive condition that currently has no cure, except in some instances of operable CTEPH.

Targeted Therapies for Pulmonary Arterial Hypertension

Oral Medications

Endothelin Receptor Antagonists (ERA)
- Ambrisentan (Volibris or generic)
- Bosentan (Tracleer or generic)
- Macitentan (Opsumit)
- Macitentan + Tadalafil (Opsyvni)

Phosphodiesterase Type-5 Inhibitors (PDE-5)
- Sildenafil (Revatio or generic)
- Tadalafil (AdCirca or generic)
- Macitentan + Tadalafil (Opsyvni)

Soluble Guanylate Cyclase Stimulator
- Riociguat (Adempas)

Selective IP Receptor Stimulator
- Selexipag (Uptravi)

Parenteral Medications
(not by mouth/stomach)

Prostacyclin Analogues/ Prostinoids
- Epoprostenol (Flolan)
- Room temperature stable epoprostenol (Caripul)
- Treprostinil (Remodulin)

Activin Signalling Inhibitors
- Sotatercept (Winrevair)

Intravenous prostacyclin analogues are delivered continuously through a catheter (tube) inserted and tunnelled into a vein in the chest and then infused by a portable infusion pump. All prostacyclin analogues are available intravenously.

Subcutaneous prostacyclin analogues are delivered continuously through a needle, most often inserted under the skin of the abdomen (belly) and then infused by a portable infusion pump. Treprostinil is the only prostacyclin analogue available subcutaneously.

Treprostinil may also be delivered through an inhaler. Inhaled medications for pulmonary hypertension are not currently available in Canada.

Sotatercept is delivered as an injection once every three weeks.

Learn more about the types of treatment options available to you at
phacanada.ca/treatmentoptions

Additional Treatments

Doctors might also prescribe non-PH-specific medication, such as:

Calcium Channel Blockers (CCB)

CCBs are blood pressure medicines used for systemic hypertension. CCBs can decrease pulmonary artery pressures in some PH patients who are “vasoresponders” to certain medications given during their right heart catheterization.

Diuretics

A diuretic - sometimes called a “water pill” or “fluid pill” – is a chemical that helps you lose water by increasing the amount of urine you eliminate. Diuretics help to rid excess fluid that puts pressure on the heart, liver, digestive organs, and lower extremities.

Blood Thinners (anticoagulants)

Anticoagulants (such as heparin or warfarin) are medicines that help prevent blood clots.

Supplemental Oxygen

Low oxygen levels in the lungs can cause further narrowing of the blood vessels in your lungs, making it harder for blood to flow through your lungs. Your PH team might call this “an increase in resistance.” Supplemental oxygen can help some patients by relaxing the blood vessels in the lungs, which can improve how you feel.

If prescribed by your PH specialist, supplemental oxygen use is just as important as the medications you are prescribed. There are many different oxygen device options. Your healthcare team will work to balance oxygen flow and portability depending on your medical needs and insurance coverage.

Treatment Pathways

Changes in body chemistry can have a profound impact on overall health. A change in chemical balance affects how cells communicate with each other. That imbalance can cause cells and organs to react abnormally.

Four chemical families or pathways are known to be abnormal in the blood vessels of the lungs of PAH patients. These abnormalities contribute to PAH by causing the blood vessels in the lungs to tighten.

PH-targeted medications work by modifying the chemical imbalance in these pathways. They help blood vessels relax so the blood flows more easily through them to the lungs.

Sotatercept: a new treatment pathway for pulmonary arterial hypertension

In August 2024, Health Canada approved sotatercept (Winrevair), a medication in a new class of treatments known as activin signalling inhibitors, for pulmonary arterial hypertension patients in functional classes II and III (see page 24). As of December 2024, it is not yet available for routine use in Canada and is not covered by most private insurance or public drug plans.

Sotatercept works by blocking cell overproduction and remodelling in the pulmonary arteries. Certain cellular signals called activins stimulate abnormal cell growth. Blocking certain activins improves the balance between pro-proliferative and anti-proliferative signalling, resulting in less abnormal cell proliferation and lower pressure in the pulmonary arteries.

As someone who has experienced all 3 prostacylin medications, I can tell you that these are some of the most efficient treatments that help stabilize PH and offer the patient a semi-normal quality of life.

Nicole Dempsey
Living with PAH since 2013
Cambridge, ON

1 Endothelin pathway

Too much endothelin-1

Endothelin is a family of chemicals made by the cells in the innermost layer of the blood vessels. Endothelin can tell the blood vessels to relax or tighten.

One form of endothelin, called ET-1, tells the blood vessels to tighten by binding to a specific receptor on a nearby cell's surface. It can be thought of as a key (ET-1) fitting into a lock (receptor).

Many PH patients make too much ET-1. Endothelin receptor antagonists (ERA) block the receptors from the increased ET-1, decreasing its effects and allowing the blood vessels to relax and widen.

2 Nitric oxide pathway

Too little nitric oxide

Nitric oxide is a gas naturally produced by the body that circulates in the blood. Nitric oxide acts as a key for a lock called soluble guanylyl cyclase (sGC). sGC can be thought of as a factory (called an enzyme) that produces an important chemical called cyclic GMP (cGMP).

Cyclic GMP is an important chemical because it helps blood vessels relax. It is broken down by another enzyme called phosphodiesterase type-5 (PDE-5). Many patients don't have enough nitric oxide to fuel the production of cGMP.

PH-targeted treatments help increase cGMP and keep blood vessels more relaxed in two ways. The first is PDE-5 inhibitors, which stop PDE-5 from breaking down the important cGMP. The second is with a sGC stimulator (or agonist), which makes the sGC factory produce cGMP with less fuel (nitric oxide).

3 Prostacyclin pathway

Too little prostacyclin

Prostacyclin is also produced in the cells of the innermost layer of blood vessels. It also helps those blood vessels relax and widen. Prostacyclin also can be considered a key that fits into a lock called the prostacyclin IP receptor.

Many PH patients don't make enough prostacyclin. PH-targeted treatments help increase prostacyclin in two ways. The first is prostacyclin analogs (or prostinoids), synthetic forms of prostacyclin that unlock prostacyclin IP receptors. The second are prostacyclin IP receptor stimulators (currently only selexipag). The receptor stimulators make prostacyclin IP receptors more sensitive to prostacyclin, so they are easier to unlock with prostacyclin chemicals.

Learn more about the pathways used to treat PH at
phacanada.ca/treatment-pathways

Functional Classes

Doctors rate the severity of PH symptoms through four functional classes. The classes describe the severity of a patient's symptoms and can help doctors determine treatment options. Many people fall under higher classes at diagnosis because they aren't diagnosed in the early stages of PH. However, one goal of treatment is to move into a lower functional class. While many people receive a difficult prognosis (the likely course of a disease) initially, many find their health improves with treatment and self-management.

The following descriptions are modified from World Health Organization functional assessments for pulmonary hypertension.

Class I

You have no symptoms during ordinary physical activity.

Class II

You are comfortable at rest, but your ordinary physical activity causes some breathlessness, chest pain, fatigue, or dizziness.

Class III

You usually have no symptoms at rest, but breathlessness, chest pain, fatigue, or dizziness greatly limits routine activity.

Class IV

You are often breathless and tired, even while resting. You can't do any physical activity without symptoms, or you faint with activity.

As your functional class or risk status changes, your doctor might change the type, amount, or delivery method of your medication.

Finding a PH Centre

Centres specializing in the treatment of PAH and CTEPH are located throughout Canada. Their experienced medical teams include PH nurses and specialists such as cardiologists, respirologists, and surgeons.

When is it important to see a PH specialist?

Significant progress has been made in the landscape of PH diagnosis and management in Canada, however prognosis remains poor for many patients and challenges remain. Referral to a PH centre of excellence is essential when a diagnosis of PAH or CTEPH is suspected, or when the cause of PH cannot be confirmed with initial investigations.

Please note that some provinces require patients to be followed by a specialist to receive coverage for PH-specific medications.

To find a clinic near you, please visit the "Find a PH Centre" directory at **phacanada.ca/phcentres**

The information provided on this page is for informational purposes only. PH Centres in Canada are not formally accredited by PHA Canada.

TIP: Keep a list of questions between doctor visits. Don't be afraid to ask why your doctor is running a test, scheduling a procedure, or recommending a change in medication.

Preparing for Medical Appointments

Preparing for your medical appointments is an important step in taking charge of your treatment plan. Being prepared for visits with your specialists can help you get the most out of them, reduce stress, and establish effective communication with your medical team. Here are some suggestions for preparing for appointments.

Learn about pulmonary hypertension

At any stage of your journey, it can be empowering to understand what's happening to your body and learn about your treatment options so you can take an active role in managing your disease. Ask your medical team about which information is relevant to you and if they can provide resources.

PHA Canada has many print and electronic resources available to help educate patients and caregivers:

- PHA Canada's website provides comprehensive information to help patients and caregivers better understand PH, including its causes, diagnosis, and treatment: **phacanada.ca**
- Information and resources for CTEPH patients: **phacanada.ca/CTEPH**
- Information and resources for families of children with PH: **phacanada.ca/pediatrics**

Fill out paperwork ahead of time

Call the office and ask them to email or mail you any paperwork you will be asked to fill out. This way, you will have time to fill out forms at home and provide all necessary details. Make two copies; keep one for your own records, and bring the other to your appointment.

If you cannot get the paperwork in advance, write down your medical history and relevant information (such as a list of medications) before your appointment. Preparing this information ahead of time will make it easier to fill out forms at the clinic and will help you remember key facts.

Have someone accompany you to your appointment

When possible, it is a good idea for your primary caregiver to accompany you to your appointments. They can provide you with moral support during the visit and be there to ask questions. After your appointment, they can help you remember information provided by your medical team. Being present at appointments will also help your caregiver understand your disease better and empower them to advocate for you in emergency situations.

Create a file for all your medical information

Organizing your medical information in a file (or binder) can help you keep track of your treatment plan and the evolution of your symptoms. You can take this file to all your appointments and ask doctors for copies of your medical records.

Your medical information file should include the following:

- A list of all your doctors and their contact information;
- A list of your medications (including the name of medication, dose, and frequency, why you take the medication, when you began taking it, and which physician prescribed the medication). If possible, bring medications in their original containers to your appointments;
- A copy of all your medical records;
- Copies of test and lab results; and
- A pad of paper to write down questions and take notes.

Make a list of questions

Doctor's appointments can be overwhelming, and it's easy to forget to ask an important question. Writing down questions for your medical team can help ensure you remember them for your next appointment. As questions come up in your day-to-day life, jot them down in your medical file; that way, you will be sure to ask them at your next appointment.

Questions you may want to ask:

- How often should I come to see you?
- What changes should I make to my diet, exercise routine, or lifestyle?
- What does this test/procedure involve?
- What will the results of this test/procedure tell you?
- What are the potential complications/risks of this test?
- How will this medication improve my health?
- What are the potential side effects of this medication?
- What resources are available to minimize pain and other side effects?
- What should I be preparing for in the future?

Do not be afraid to ask your medical team for further explanation if you don't understand the information they provide. Remember that communicating effectively with your medical team helps ensure that you are receiving the best possible care.

You are your own best advocate

Although members of your medical team are experts in PH, you know your body best. Don't be afraid to ask for explanations and tell your doctor or nurse about your unique circumstances. By understanding your individualized condition and personal treatment goals and communicating effectively with your medical team, you will be better able to meet your needs.

Living With PH

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Impact of Diagnosis

Every new patient responds differently when learning that they have PH. You might feel angry, frightened, lonely, frustrated, overwhelmed, worried, numb, or a combination of feelings.

Remember: There are no right or wrong reactions. Your feelings are normal, and you aren't alone. Treat yourself with kindness and compassion by allowing yourself to feel and respond to your diagnosis without judgment.

Medical treatment is only one component of caring for yourself. You must also work through your emotions and rediscover meaning and purpose in your life. The emotional and mental components of your health need time and attention to allow you to feel your best.

Download PHA Canada's Emotional Wellness Handbook to help you work through the emotional impacts of your diagnosis at **phacanada.ca/resources**

Upon diagnosis, I decided to measure life in quality rather than quantity. For me, this means finding joy and happiness each day - an ice cream cone, my favourite blanket, a groan-worthy dad joke, a road trip to nowhere, and hugs from my boys. I remind myself that, in life, the little things are the big things, which means that even on the darkest days, small rays of sunshine can offer a reprieve from the heaviness that we carry.

Angèle Belliveau, Living with PAH since 2021, Dieppe, NB

While it can be difficult to imagine in the days and months following diagnosis, many patients develop strategies to cope with the physical and emotional aspects of living with PH and lead happy, fulfilling lives. With a bit of patience, planning, and flexibility, many people affected by PH find ways to work, travel, exercise, and do the things they loved before they were diagnosed.

Pregnancy

Pregnancy is generally considered unsafe in patients with PH as it is associated with high rates of maternal deaths and should be avoided. Pregnancy places very high demands on the heart, and when affected by PH, the heart cannot keep up. In pregnancy, people with PH can experience miscarriage, low birth weight babies, heart failure, and even death, often around the time of delivery.

Your PH team will speak to you about safe and effective methods to avoid pregnancy. They can also help refer you to surrogacy and adoption options, which are safer options for most patients with PH who desire children. Many patients are diagnosed with PH during or shortly after pregnancy. It is thought that the hormonal and blood flow changes in pregnancy may uncover the disease in these cases.

Additionally, some PH-targeted therapies and some other medications used in PH clinics are known to cause birth defects in infants (known as teratogenic medications).

PH patients should speak to their PH team about the safety of pregnancy before becoming pregnant.

Sexual Health

PH can change the way you view yourself and your body, affecting your sense of self, body image, and intimate relationships.

Changes to your health and body can be difficult to adjust to and can make sexual relationships and reproductive decisions more complicated. Physical symptoms such as fatigue, breathlessness, weight fluctuations, and medication side effects can affect the intimacy of people with PH. Medication pumps, tubing, infusion sites on the skin, as well as a nasal cannula and oxygen tanks, may also pose challenges to engaging in sexual activities.

A patient's sexual partner may also have fears about hurting them during intercourse, possibly worsening their health conditions, or potentially causing pregnancy.

Sexual intercourse and intimacy are often safe when living with PH. However, it is recommended you discuss your individual circumstances with your medical team. Talking openly with your sexual partner(s) about your fears and concerns will also help make physical intimacy easier.

Ways to Cope

Find Community Support – Connecting with others with PH is a great way to get answers to specific questions or help you come to terms with your diagnosis. PHA Canada can help connect you to the support you need. Visit **phacanada.ca/support** to find the support that's right for you and your family.

Seek Professional Help – Treating your mental health is as important as treating your physical health. Untreated depression and anxiety can affect your physical and emotional health and interfere with your ability to care for yourself or others. Consult your PH care team or primary care provider about medication, counselling, and other treatment options. Your healthcare team might refer you to a psychiatrist, social worker, or other mental health professional.

Take Care of Yourself – Whether you are a newly diagnosed patient or a caregiver, journaling, meditating, eating well, and exercising can reduce stress and improve mental and physical health. For more ideas on coping, exercise, and day-to-day living, visit **phacanada.ca/resources**

Seek Accommodations – Changing the way you approach work (including school) can help you manage your symptoms, side effects, and medical appointments while maintaining a job. You may be entitled to adaptations to your work/school schedule or environment to help reduce barriers to your ability to perform.

Explore Government Assistance – Many people with PH will qualify for government tax benefits and financial support programs that support people with disabilities or chronic health conditions. Consider applying for programs such as the Disability Tax Credit or an accessible parking permit. For more information on government assistance programs, visit **phacanada.ca/finance**. You may also consider speaking with a social worker.

Get Involved – There are many ways to get involved in the PH community. By sharing your experience with PH, you can help raise awareness, educate people about PH, and provide hope and support to others. Find local and virtual events, volunteer opportunities, and more at **phacanada.ca**

Nutrition, Exercise, and Energy

How you manage your diet, exercise, and energy is an important aspect of your care plan. Your care team may ask you to limit your sodium intake, help you create an exercise plan, and provide tips to conserve your energy.

Sodium and Fluid Retention

Your PH care team will likely recommend dietary changes to reduce PH symptoms and improve heart health. One of the most important recommendations is a low-sodium diet. Sodium causes fluid retention, which can worsen symptoms and put more pressure on the heart – a serious risk for people with PH.

A buildup of excess fluid or swelling is called edema. You will notice it when your legs, ankles, or abdomen swell, and it will show up as weight gain. It can be helpful to weigh yourself daily and track any changes in your symptoms. Weigh yourself at the same time every day with the same scale. The best time to weigh yourself is in the morning after using the bathroom. Ask your care team when to notify them that your weight is increasing.

Sodium causes fluid retention, which can worsen symptoms and put more pressure on the heart.

Limiting Sodium

Sodium is a naturally occurring element in many foods, especially table salt. That's why many people with heart issues ask to "hold the salt" when eating away from home. At home, you can reduce sodium intake by not adding salt when cooking, avoiding salty snacks, and checking nutrition labels on prepared foods.

Instead of using salt, try:

- Lemons
- Limes
- Garlic
- Other herbs and spices

Find more about nutrition and PH, including a low-sodium recipe book at **phacanada.ca/nutrition**

How much exercise is right for you?

Regular exercise can improve cardiovascular capacity (the ability of the body to take in oxygen and deliver it to your muscles and organs), muscle function, and quality of life for people with PH. Even a small amount of exercise, such as walking slowly to the opposite side of the room, can have positive health benefits.

Exercise recommendations differ for each patient based on the severity of PH and other health factors. Those recommendations might change over time depending on symptoms and response to treatment.

Always talk with your PH care team before beginning an exercise program. Your PH specialist might recommend cardiac or pulmonary rehabilitation or help you create an exercise plan that works for you.

Maximize Your Energy

Finding the right balance of activity and rest can be challenging for people with PH. You know better than anyone how much PH can drain your energy and affect your mobility. Sometimes, even a walk is out of the question.

Not being able to do the things you did before PH isn't a sign of failure. Because of your PH, you need to be more selective about what you can do and what's important to you. Take time to re-examine your priorities.

Consider these strategies to conserve your energy:

Home and Office

- Set a timer during activities to remind yourself to take a break. For example, if you have physical work to do, set a timer for 10-15 minutes. When the timer goes off, stop working, hydrate, and rest for at least one minute for every minute worked.
- Keep anything you use often at hip level. Avoid stooping, crouching, or bending. If you must bend, get up slowly.

Lifestyle

- Get comfortable saying no and not feeling guilty about it. If you tend to wear yourself out by helping other people, take a hard look at how you're doing before accepting requests, invitations, or activities.
- Take breaks or naps as often as you need.

On the Go

- Obtain a parking permit for persons with impaired mobility. Ask your doctor for certification to send to your provincial or local department of transportation.
- Use mobility devices in places that require a lot of walking. Save your energy to enjoy the experience.

Find more tips about exercise and PH at **phacanada.ca/exercise**

Support for Caregivers

From the moment your loved one was diagnosed with PH, your life changed. Chances are, as your shared journey with PH continues, you may also take on the role of a "caregiver."

As a caregiver, you'll spend a lot of time caring for the person with PH. This can be emotionally or physically exhausting, putting your own health at risk. It's important to prioritize your well-being, not just for yourself but also for your loved one with PH.

Watch for these common signs of caregiver stress:

- Feeling overwhelmed or constantly worried
- Feeling tired often
- Getting too much sleep or not enough sleep
- Gaining or losing weight
- Becoming easily irritated or angry
- Losing interest in activities you used to enjoy
- Feeling sad
- Having frequent headaches, bodily pain, or other physical problems
- Abusing alcohol or drugs, including prescription medications

Take the time to recognize the signs of burnout in yourself and plan for how you can prioritize your needs. Just like those who are newly diagnosed, you – as a caregiver – must also find time to care for yourself and focus on activities or techniques to reduce stress.

Organizations that support caregivers are available across Canada, offering support, resources, and tools at the local, provincial, and federal levels. Find a list of organizations near you at **phacanada.ca/caregivers**

I am looking forward to spending the rest of my days watching our beautiful little girl grow into a lovely woman.

Jerry Clark
PAH Caregiver since 2019
Surrey, BC

Preparing for Emergencies

Collaborate with your support network to devise a tailored emergency plan that considers the severity of your PH, location, support systems, and personal factors. Share this plan with your PH care team for effective preparedness.

Here are a few tips to get started:

- Sign up for emergency alerts.
- Stock up on necessities, especially drinking water, food, first aid kits, flashlights, and batteries.
- Keep a supply of extra distilled water for CPAP machines.
- Ask your PH care team if you can get extra medication and/or supplies (cassettes, syringes, etc.). Some insurance companies will allow early refills for emergencies or disasters.
- When seeking emergency medical care, bring a 4–5-day supply of your PH medications with you, as they may not be available from the hospital.
- Keep your insurance cards with you when you leave home.
- Always keep a list of all your medications with you. Infusion patients should state that their infused medication should never be interrupted or discontinued.
- If you have a central IV line (catheter), keep cleaning and dressing supplies with you, including gloves, masks, alcohol pads, dressings, and tape.
- If you use oxygen, ask your oxygen company to have your tanks filled so you don't run short. Also, ask about extra batteries.
- Contact your power company about your health condition in case they can list you as a high priority in a power outage. Work with your health provider to complete/submit paperwork to ensure your power stays on in an outage.
- Contact your specialty pharmacy if you're running low on medication, need to change your medication shipment address, or have other therapy-access concerns.
- If you are a member of a local support group, exchange phone numbers with the leader and other members to keep in touch during unexpected events.

Find tools at **phacanada.ca/emergencies**

If you experience any of these symptoms or situations, seek emergency help or dial 911:

- Loss of consciousness
- Coughing up blood
- Pump has stopped
- You run out of medication for your pump
- Dislodged line/damaged or non-functioning catheter

Keep a list of numbers with you for your specialty pharmacy, PH doctor/clinic, and emergency contacts.

Canadian PH Community

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About PHA Canada

PHA Canada (Pulmonary Hypertension Association of Canada) is a federally registered charity established in 2008 by patients, caregivers, and healthcare professionals to create a better life for all Canadians affected by pulmonary hypertension and represent a united national PH community. Our mission is to empower the Canadian pulmonary hypertension community through support, education, advocacy, awareness, and research.

> Learn about PHA Canada's tradition of advocacy, support, and awareness at **phacanada.ca/history**

PHA Canada is committed to working with and for all Canadians affected by pulmonary hypertension and is proud to be the national voice of Canada's PH community. True to the vision of our founders, PHA Canada strives to represent and engage those affected by PH in every corner of the country so that you feel heard, informed, and supported no matter where you live.

PHA Canada exists today because of the hard work and dedication of people just like you. Get inspired by the stories of the "Eternal PHriends of PHA Canada" at **phacanada.ca/eternalphriends**

Get to know the PHA Canada team! Located throughout the country, our staff team is here to support you through every step of your journey. Learn more at **phacanada.ca/staff**

Support & Education Resources

Like most people recently diagnosed with PH - or close to someone newly diagnosed - you have probably never met anyone else with the disease. It can be a huge relief to connect with others affected by PH, share experiences, and understand that you're not alone. Many people have gone through similar journeys and understand what you're going through. And they want to help.

PHA Canada helps to facilitate a robust network of support across Canada to ensure no one affected by PH ever feels alone. We are here to provide support, hope, and resources to help patients and their loved ones adjust to life with PH. Here are some of the resources available:

Support

- Support groups are local and online groups where patients, families, friends, and healthcare providers come together to talk about PH. Being part of a support group (whether in-person or online) can help relieve the stress and isolation that naturally comes with living with a rare disease. It can also help you learn more about how to manage life with PH.
- The PH Buddy Program matches you with a peer from the PH community who can understand what you are going through and help provide support and guidance. Your 'Buddy' can support you via email, phone, or even in person. This program is for patients, caregivers, or any friend or family member affected by PH.
- Community Meet-ups are virtual drop-in spaces where the PH community meets over Zoom. Meet-ups are not support groups—they are open, social gatherings where patients and caregivers can chat with others from the PH community.

Find details about these support services at **phacanada.ca/support**

Education

PHA Canada's website features a variety of educational materials and videos, including:

- Information about the different types of PH
- Resources for living well with PH, including on mental health, vaccination, nutrition, and exercise
- Information about associated conditions such as COPD and scleroderma
- Oral medication information sheets
- Tools for navigating complex issues such as school, work, contraception, intimacy, and end-of-life planning

Find resources at **phacanada.ca/resources**

Community Conference

PHA Canada is committed to bringing the PH community together to learn more about PH and make valuable connections with one another. Every year, we support patients, caregivers, and medical professionals to get together to share their knowledge and experiences.

View sessions from previous conferences at **phacanada.ca/conference**

Advocacy & Awareness

PH Awareness Month and World PH Day are two significant events that bring attention to pulmonary hypertension (PH). PH Awareness Month takes place in November and is a dedicated time to raise awareness about PH and encourage early diagnosis. It's a month full of events, information, and support. World PH Day, observed on May 5th each year, is a global initiative to unite the PH community, emphasize the importance of PH awareness worldwide, and promote understanding and research. These events provide opportunities for patients and their families to connect, share experiences, and advocate for better care and treatments. They play a vital role in ensuring that no one faces PH alone.

Learn how you can help raise awareness of PH at **phacanada.ca/awareness**

Stay Connected

Join our vibrant community and stay at the forefront of what matters most! Subscribe to "The Pulse," our Monthly Newsletter, for the latest updates, insights, and exclusive content delivered straight to your inbox every month. Plus, dive deeper into inspiring stories, in-depth articles, and community events with our biannual magazine, "Connections." Don't miss out on the chance to be informed, inspired, and connected – subscribe now and be part of our journey!

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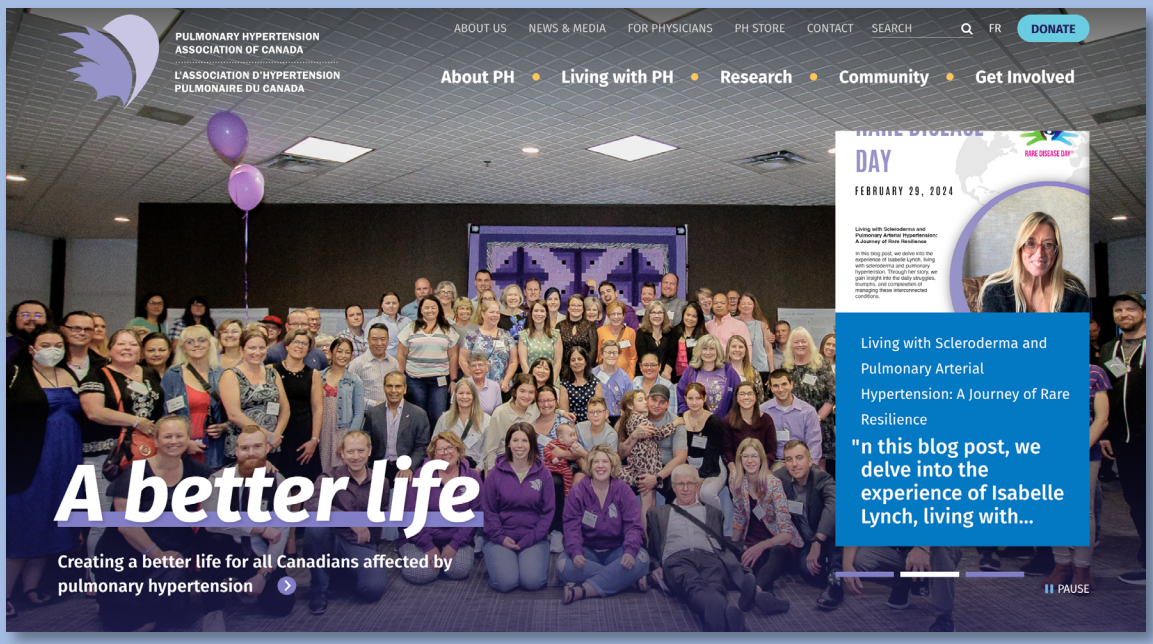

Participate in Research

Scientists and clinicians need patients and caregivers as partners in research studies and clinical trials. No one knows PH like those who live with it every day. You can provide insight about PH that researchers and physicians do not have. Every treatment that exists today is the direct result of individuals who have stepped up to volunteer for research.

Not all research studies test a new treatment. Some studies look at the safety or efficacy of a new drug or therapy, while others may collect information to help us understand the impact PH has on patients' lives and how we can improve quality of life.

Canadian Pulmonary Hypertension Registry (CPHR)

The Canadian Pulmonary Hypertension Registry (CPHR) is a database of patient information collected to help better understand PH and improve patient care. This registry allows us to understand PH better and its impacts on patients. Your local PH team will have more information about the registry and how to sign up.

Learn more about the Canadian PH Registry at **phacanada.ca/CPHR**

Clinical Trials in Canada

Patient participation in research is essential to making scientific advancements in the diagnosis and treatment of PH. Numerous early-stage and clinical-stage research studies are currently taking place in research centres throughout Canada.

Learn more about Clinical Trials in Canada at **phacanada.ca/clinicalresearch**

Non-Clinical Research Studies

Learn more about non-clinical research studies currently seeking participants, as well as results and recommendations from previous research projects involving the PH community at **phacanada.ca/survey**

Talk with your PH team about research opportunities.

Notes:

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Notes:

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Creating a better life for Canadians affected by PH.