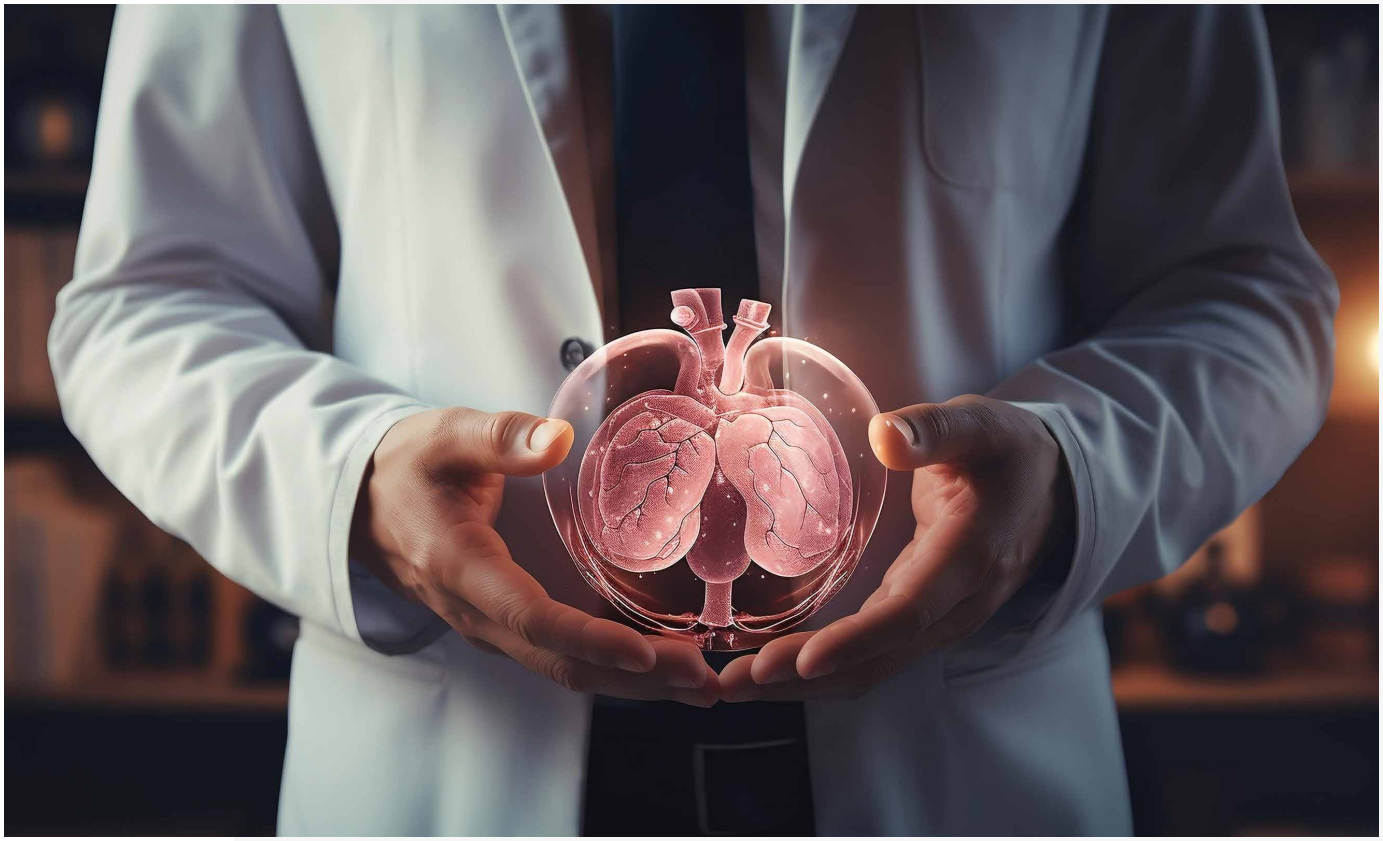
RESPIPLUS™ PRESENTS

JULY 2024

A CANADIAN INITIATIVE: EXPLORING ADULT PULMONARY HYPERTENSION



RESPIPLUS

A CANADIAN INITIATIVE: EXPLORING ADULT PULMONARY HYPERTENSION

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The contents of this report are not to be used as guidelines and should not influence any patient treatment plans. The following is based on the opinions of the expert panel members who have an interest in pulmonary hypertension and, more specifically, pulmonary arterial hypertension in adults. It is for informational purposes only. It is not intended to provide or supersede medical or professional advice. Medical advice should be sought from a qualified healthcare professional for any questions. Reliance on any information in this report is solely at your own risk. We do not assume any responsibility or legal liability for the accuracy, completeness, timeliness, or quality of any information in this report.

In order to maintain anonymity and protect the privacy of individuals interviewed during this process, some names and identifying details have been changed.

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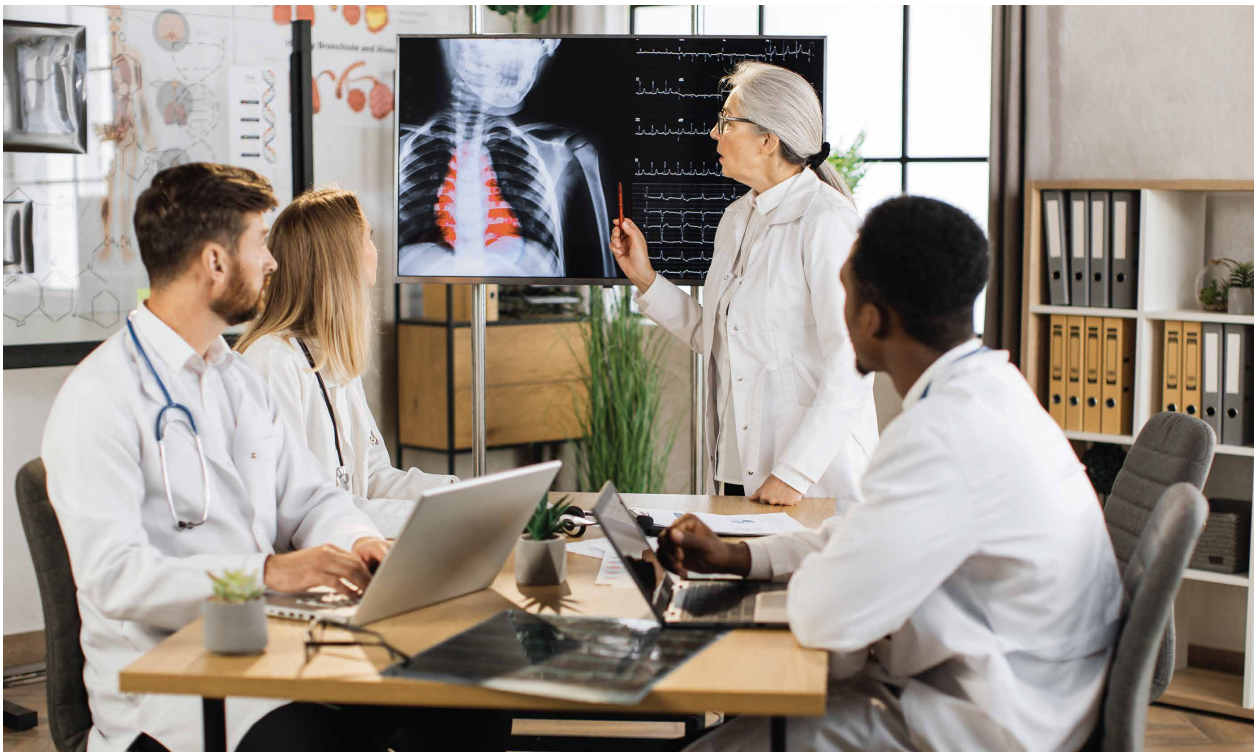
This report is available on the website www.chroniclungdiseases.com

TABLE OF CONTENTS

ABOUT THE PH REPORT



Pulmonary hypertension (PH) is a potentially life-threatening condition characterized by elevated pressure in the pulmonary arteries, leading to increased workload on the right side of the heart. It can result from various underlying causes, including but not limited to idiopathic pulmonary arterial hypertension, rheumatological diseases, liver diseases, left heart diseases, lung diseases, and chronic thromboembolic disease. Healthcare professionals should be aware of the complexities around diagnosis, management, and referral pathways. Collaboration among healthcare providers is crucial for effective PH management.

This Pulmonary Hypertension Report was prepared and published by RESPIPLUS, a non-profit organization with more than 20 years expertise in developing self-management programs for patients in the field of respiratory medicine and was overseen by a panel of clinicians, academics, healthcare professionals and patients in the field of pulmonary hypertension.

The objective of this report is to serve as a comprehensive tool tailored for healthcare professionals, patients, and the broader public in the Canadian context. It provides clear and concise information on PH diagnosis, treatment, and management, grounded in evidence-based practices. For healthcare professionals, the report offers valuable insights into diagnostic approaches, treatment modalities, and management strategies specific to the Canadian healthcare system. Similarly, for patients, it serves as an accessible resource, empowering them with broader knowledge about their condition, treatment options, and lifestyle adjustments.

RESPIPLUS, in collaboration with the Pulmonary Hypertension Association Canada (PHA Canada), the Canadian Thoracic Society (CTS) and the Family Physician Airways Group of Canada (FPAGC) have implemented a panel of key opinion leaders and scientific experts, to develop this report.

Additionally, our report emphasizes the importance of multidisciplinary collaboration in addressing the diverse needs of PH patients, promoting a holistic approach to care. This report includes evidence-based recommendations, and it also supports advocacy for policy changes aimed at improving infrastructure, access to specialized care, and research initiatives related to PH.

Finally, the report provides a platform for industry engagement, allowing stakeholders to showcase their contributions to PH research and development in the Canadian healthcare landscape.

TEAM

Panel, collaborating organizations & RESPIPLUS team members

SCIENTIFIC PANEL

Jamie Myrah, BA | Executive Director, PHA Canada | Panel Lead
Carolyn Doyle-Cox, RN, MSN, APN | Advanced Practice Nurse
Paul Hernandez, MDCM, FRCPC | Respirologist | CTS Representative
Alan Kaplan, MD, CCFP(EM), FCFP, CPC(HC) | Family Physician | Chair, FPAGC
David Langleben, MD, FRCPC| Cardiologist
Lisa Mielniczuk, BSc, MSc, MD, FRCPC | Cardiologist
Beth Slaunwhite, credentials | Patient Partner
Jason Weatherald, MD, MSc, FRCPC | Respirologist

PHA Canada

The Pulmonary Hypertension Association of Canada (PHA Canada) is a non-profit organization committed to supporting individuals affected by pulmonary hypertension (PH) and raising awareness about this rare and life-threatening disease. Through advocacy, research, education, and support programs, PHA Canada strives to improve the lives of PH patients and their families across Canada.

FPAGC

The Family Physician Airways Group of Canada is committed to helping those with airway diseases lead a full life. The group is dedicated to helping all family physicians and primary care providers maintain and increase their skill in assisting those with airway diseases like asthma and COPD. The strategy of the group is to maintain a speaker bank, a data bank, and practical tools to help physicians and primary care providers attain these skills. They have brought the primary care perspective to this investigation.

CTS

The Canadian Thoracic Society (CTS) is the national interdisciplinary medical organization. A non-profit membership association representing specialists, physicians, researchers, scientists, and respiratory healthcare professionals. CTS promotes respiratory health by enhancing the ability of healthcare professionals through best practice guidance, leadership, collaboration, research, education, and advocacy. CTS: Advancing Knowledge. Improving Outcomes.

RESPIPLUS TEAM

RESPIPLUS is a Canadian non-profit organization, with the mission of developing training and educational programs in chronic respiratory diseases of the highest level of quality, based on the most recent knowledge supported by the scientific literature and guidelines. RESPIPLUS conducts comprehensive investigations: by integrating diverse expertise and advanced methodologies, they aim to deepen understanding, improve diagnosis, and enhance treatment strategies for chronic lung conditions, ultimately advancing respiratory health outcomes.

Maria Sedeno, MM, BEng | Executive Director
Emily Horvat, BSc | Senior Project Manager
Katrina Metz, RRT | Consultant

EXECUTIVE SUMMARY

It is important to note that while our investigative endeavors focused on Pulmonary Arterial Hypertension (PAH), it would have been remiss for us not to investigate Pulmonary Hypertension (PH) as a whole. PAH and PH may sound similar, but they denote distinct conditions. While PH is common and encompasses various diseases affecting the pulmonary arteries, PAH is rarer and specifically refers to a subset characterized by narrowing of the small arteries in the lungs. It is crucial to understand their differences for accurate diagnosis and treatment. Therefore, our report not only examines PAH but also discusses the broader spectrum of PH, providing insights into both the general condition and its specific etiologies for a more comprehensive approach.

Pulmonary arterial hypertension (PAH) is a rare and life-threatening condition that poses significant challenges in terms of diagnosis, treatment, and management. In Canada, there are notable care gaps in the provision of comprehensive care and diagnosis for PAH patients.

To overcome these gaps, we carried out a comprehensive needs assessment between September 2023 and February 2024. A Scientific Panel led this project to identify areas requiring improvement, such as timely diagnosis, access to specialized care, and ongoing education for healthcare professionals. With these results, healthcare systems can work towards bridging these gaps and enhancing the quality of care provided to PAH patients across the country.

Pulmonary arterial hypertension is an uncommon cause of dyspnea and it's something that most primary care physicians and even specialists are not going to see very often. But, it has to be considered in the population of people that are short of breath where otherwise, we don't have a straight answer for why they're short of breath.

Alan Kaplan, Family Physician

When evaluating unexplained dyspnea in patients, you must think of PH as a possibility and consider ordering an echocardiogram.

Paul Hernandez, Respirologist

INTRODUCTION

Pulmonary hypertension (PH) is a potentially life-threatening vascular disorder characterized by elevated pressure in the pulmonary arteries. Its symptoms, ranging from breathlessness, fatigue, swollen ankles and legs, chest pain, bluish lips, hands and legs, dizziness with activity and fainting often mimic common cardiac or respiratory conditions, complicating the diagnosis. The global burden of PH is substantial. It is estimated to affect 1% of the world's population and increasing to 10% in those 65 years and older (most commonly due to left heart or lung disease[1]).

Within the Canadian healthcare landscape, the diagnosis, treatment, and management of pulmonary hypertension (PH) present unique challenges. There are hurdles in accurately and timely diagnosing PH, intricacies of treatment options, and the ongoing challenges in effective long-term management. Understanding these complexities is crucial in improving the outlook for individuals grappling with PH within the Canadian healthcare system.

HOW RARE IS PAH?

> **When you hear footsteps in the distance, you think horses, not zebras, right? And certainly not unicorns. And pulmonary (arterial) hypertension would be a unicorn here. Pulmonary arterial hypertension is an uncommon cause of shortness of breath and it's something that most primary care doctors and even specialists are not going to see very often.**
>
> Alan Kaplan, Family physician

PATIENT EXPERIENCE

I was diagnosed with Idiopathic Pulmonary Arterial Hypertension in 2016. Since the day I was diagnosed, I have wanted to reach other patients to show that we are not alone, and to educate and advocate so others are not misdiagnosed. I remember thinking at the time that I do not want anyone to feel as alone as I do.

For many years before I was diagnosed, I had been telling my doctor that I struggled with shortness of breath (SOB). Playing sports, jogging and even taking a walk with others were all very difficult for me. I saw a few respirologists, but they couldn't seem to find anything wrong. Even though the Pulmonary Function Tests (PFTs) did not point to asthma, I was told it must be exercise-induced asthma because nothing else could explain it. They prescribed inhalers (puffers) which were of no real use, so I carried on and continued making excuses for my SOB. When I started feeling a few twinges of chest pain, I was sent to a cardiologist. I was given a Holter Monitor at home which he said did not show anything abnormal. Then, I had a stress test that was stopped after about 3 minutes because the attending cardiologist told me I was having an asthma attack. I tried to explain that I didn't have asthma according to all the PFTs, but I was dismissed. It was very discouraging. I decided that it is what it is, no reason for it, and no need to continue searching. I thought I was just unfit.

In July of 2014, I decided I needed to get into better shape. It was ridiculous that I was so short of breath just carrying laundry upstairs! So, I began walking our dog in the local hilly park 4 mornings a week. I was determined. The first few days I did a route, and I was able to finish in around 75 minutes, even with the hills. I met other dog walkers, and it was enjoyable. Thankfully, we stopped often to scoop, let the dogs play, and to throw the ball around. But, when I walked with an 80-year-old lady and she was able to climb the hills without stopping, I started to feel really defeated. The more often I walked the route, the worse I got. I found myself pretending to stop and admire the squirrels, the plants, and heck even the trees. She enquired one day if I was ok. I told her I was just recovering from a lung infection…. anything not to appear so out of shape. It was mortifying. I soon realized we could no longer walk together, so I switched my start time on purpose, and lost a friend. What previously took 75 minutes was now taking 90 and soon 120 minutes. Fifteen months after I began those walks, it took 2 hours and 15 minutes to do the same route.

My work and relationships were also suffering. In the summer of 2015, I went to lunch with colleagues.I was not able to keep up on the way back, so I sent them ahead. The next invite, I declined, and stayed at work. I could not to make it up 3-5 stairs without needing to sit and take a break.

That October, I was at home in my kitchen trying to get some baking done and suddenly my heart rate doubled, nausea ensued, and I knew something was terribly wrong. I called my husband home from work, and we discussed ER, but I refused. The next day at work I was turning grey going from sitting to standing, feeling very faint, and holding the walls to walk. They sent me to the ER, where they were convinced something was wrong and tried to get me a cardiology consultation. I had been having more and more light chest pain, something I had gone to the doctor's for before. The EKGs never showed anything. A cardiology consultation would take 10 days.

That Friday evening, I was sent home with a diagnosis of stable angina and instructions to return if anything changed while waiting for my consultation with a cardiologist. I was also given a prescription for metoprolol. By 2 pm Sunday I was back at my new GP's office with instructions not to drive. My "stable angina" was now unstable, and I needed to go back to the ER. In the ER, I was seen by a cardiology fellow and had EKG's done every few hours. They ordered a stress test first thing Monday morning. When the third cardiology team member came early Monday, he asked me to accompany him to another room for an exam. He basically ran there, in my opinion, but assured me he didn't. I could not follow him; I was too short of breath. He then informed me I could not have the stress test. I started to cry in that hallway, holding on to my husband. I needed that stress test; something was very wrong.

I had had a CT Angio, there were no blockages, so what else could it be? Again, I was devastated. We sat in an office and he really, really thought about it. He was a cardiology resident and suggested that perhaps an echocardiogram (echo) might help. It was a last resort. He could not think of anything else that might replace the needed but now unattainable stress test. It was that last resort echo that finally showed Pulmonary Hypertension (PH). This is why I advocate. Three cardiology-related physicians, two respirologists, several ER physicians, a GP, and the cardiologist who had requested a stress test and a Holter Monitor several years prior never considered PH. Not one of them ever requested an echo. PH was not on their radars as a potential diagnosis and I now believe it should have been, especially with the severe unexplained SOB.

I had my right heart catheterization a few months later and my therapy started that day. I've been blessed ever since. My pressures remain low and consistent, even though I still struggle with ongoing symptoms.

DEFINING PH

As defined by the 2022 ESC/ERS Guidelines2, pulmonary hypertension is characterised by a mean pulmonary artery pressure (mPAP) ≥20 mm Hg at rest. Pulmonary Hypertension (PH) is classified into five groups based on the underlying causes and attributes. Here is a brief overview of the five groups[3]:

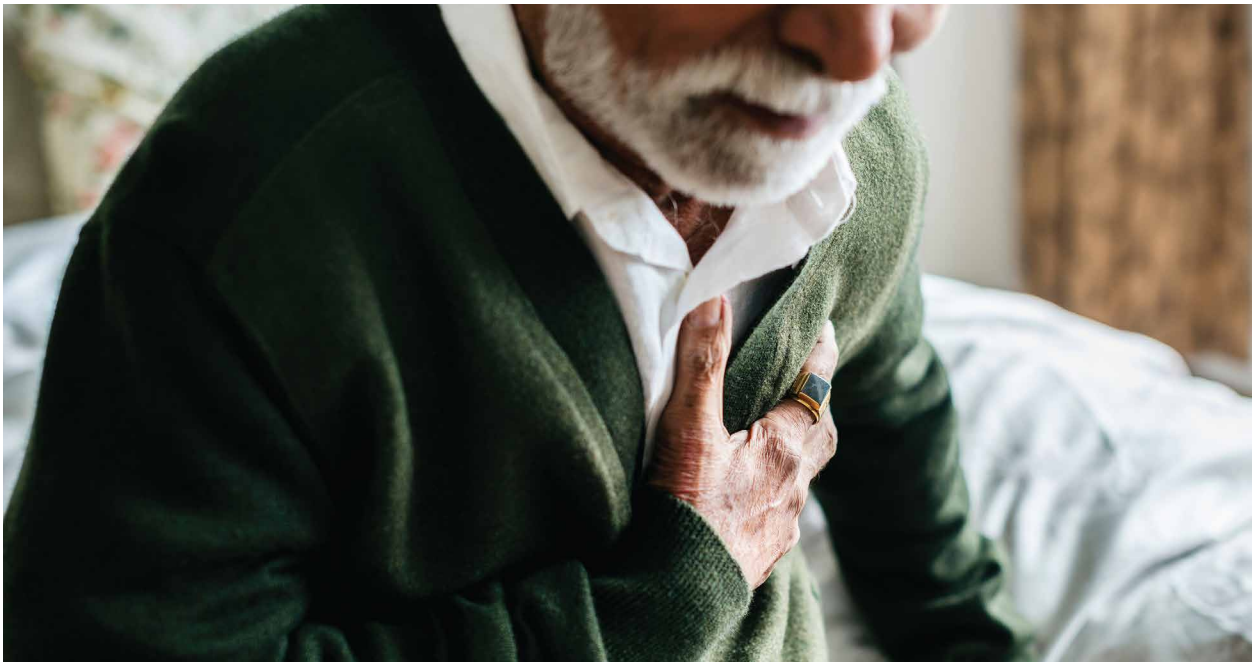
1. **Group 1: Pulmonary Arterial Hypertension (PAH):**
- Characterized by elevated blood pressure specifically in the smallest pulmonary arteries just before the capillaries, which are blocked by cellular growth that has a pattern unique to group 1.
- Includes idiopathic PAH, hereditary PAH, and PAH associated with other conditions (e.g. connective tissue disease, liver disease, HIV, congenital heart disease).

2. **Group 2: Pulmonary Hypertension due to Left Heart Disease:**
- Resulting from left-sided heart conditions such as heart failure and valvular diseases.
- Increased pressure in the pulmonary arteries is secondary to left heart dysfunction.

3. **Group 3: Pulmonary Hypertension due to lung diseases and/or Hypoxia:**
- Associated with chronic lung diseases like chronic obstructive pulmonary disease (COPD) and interstitial lung disease.
- Hypoxia (low oxygen levels) contributes to pulmonary vascular changes specifically in the pulmonary arteries.

4. **Group 4: Chronic Thromboembolic Pulmonary Hypertension (CTEPH):**
- Caused by blood clots (thromboemboli) in the pulmonary arteries.
- Not typical blood clots but chronic fibrous obstructions specifically within the pulmonary arteries that do not resolve with anticoagulants and must be physically removed (pulmonary thromboendarterectomy) or treated with a Balloon Pulmonary Angioplasty (BPA). Chronic obstruction leads to increased pressure.

5. **Group 5: Pulmonary Hypertension with Unclear or Multifactorial Mechanisms:**
- Encompasses cases where the exact cause of PH is uncertain or involves a combination of factors.
- Includes conditions such as blood disorders, systemic diseases, and metabolic disorders.



Each group has distinct features, diagnostic criteria, and treatment approaches. This classification helps guide healthcare professionals in understanding the diverse etiologies of PH and tailoring appropriate management strategies. **Because PH has nonspecific symptoms particularly in early disease, it is frequently attributed to other more common cardiac and pulmonary diseases. It is often not recognized until patients develop symptoms of right heart failure and therefore the diagnosis can take years.**

> My impression is that for certain groups of people, it's pretty long between symptom onset and diagnosis. It can be a couple of years, especially people who are older, or people who are obese. Often they have symptoms for quite awhile, but they get blamed on other things. Then they don't actually get testing of any kind until later on in the course of the disease. But there's such a large range. I think the literature suggests it's about two years from symptom onset until diagnosis, but I think there's people that have symptoms for much longer than that and maybe their symptoms weren't due to pulmonary hypertension in the first place, and they just happen to have it. And then there are people that have symptoms and they are diagnosed within a few months and it's quite rapid. So, the spectrum of wait times is quite broad from symptom onset to diagnosis for many reasons.
>
> Jason Weatherald, Respirologist

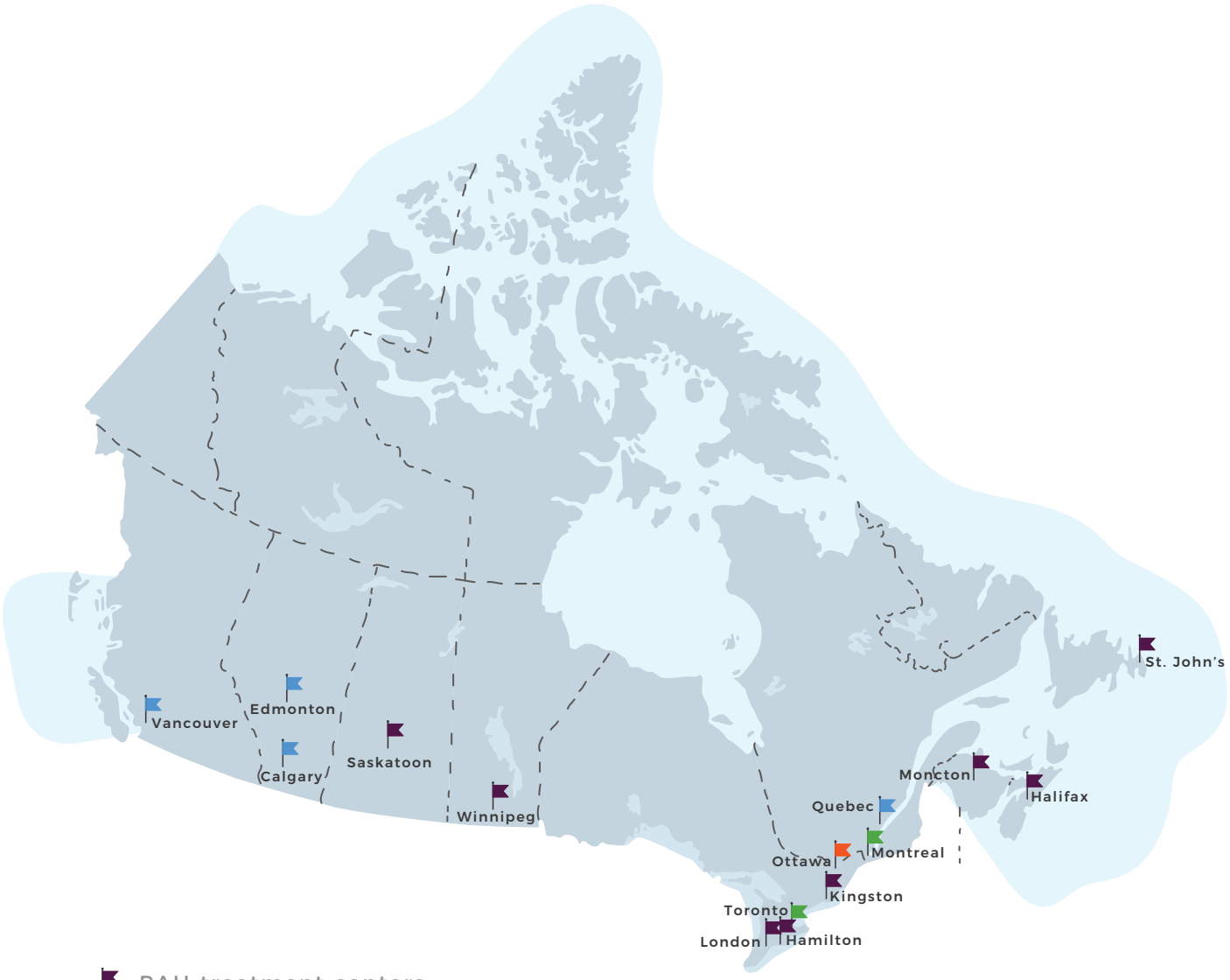
PREVALENCE AND PARTICULAR CHALLENGES IN CANADA

PH stands as a difficult challenge within the Canadian healthcare system, characterized by its increasing prevalence and complex array of obstacles. True prevalence of the disease will depend on the population being studied, and even the way PH was diagnosed (i.e. echo, catheterization, administration records..). Because of these factors, characterizing the prevalence is very challenging.

- **PH can affect anyone,** regardless of age, sex, or social/ethnic background.
- PH can affect patients not just physically, but **financially, socially and emotionally** as well.
- **Centres specialized in the treatment** of PH in adults and children are located throughout the country.
- **PAH is not a common condition.** It is estimated that approximately 2,000 Canadians have been diagnosed with pulmonary hypertension, but as many as 4,000 may be affected by the condition.
- Many people with PAH in Canada spend **two to three years seeking an accurate diagnosis.** More importantly, without treatment, the average life expectancy of a person with PAH is less than three years.
- A number of **treatments are approved in Canada** to slow progression of PAH and alleviate symptoms.
- **Disparities exist in the quality-of-care** PAH patients may receive depending on what province or territory they live in or whether they have private insurance.

CENTERS ACROSS CANADA

Below is a map of a specialized treatment centers across Canada[1,4].



- PAH treatment centers
- Adult & pediatric PAH treatment centers
- Adult & pediatric PAH and CTEPH treatment centers
- PAH and CTEPH treatment centers

More information is available at https://phacanada.ca/phcentres

ETIOLOGY AND PATHOPHYSIOLOGY

The etiology and pathophysiology of PH can vary depending on the specific type of PH:

Group 1: Pulmonary Arterial Hypertension (PAH):

- **Etiology:** The exact cause of PAH could be idiopathic or associated with other conditions such as connective tissue diseases, liver diseases, congenital heart diseases, drugs/toxins, and HIV infection. PAH could also have a genetic predisposition.
- **Pathophysiology:** PAH is characterized by narrowing and obstruction of the pulmonary arteries by remodelling and cellular dysfunction, leading to increased resistance. This results in right ventricular hypertrophy and, if untreated, can progress to right heart failure.

Group 2: Pulmonary Hypertension due to Left Heart Disease:

- **Etiology:** Caused by left-sided heart diseases such as left ventricular systolic or diastolic dysfunction, valvular heart disease, or congenital heart diseases.
- **Pathophysiology:** Increased pressure in the left atrium is transmitted to the pulmonary circulation, leading to elevated pulmonary artery pressures.

Group 3: Pulmonary Hypertension due to Lung Diseases and/or Hypoxia:

- **Etiology:** Associated with chronic lung diseases like chronic obstructive pulmonary disease (COPD), interstitial lung disease, and sleep-disordered breathing.
- **Pathophysiology:** Hypoxia, parenchymal destruction and chronic inflammation contribute to vascular remodeling in the pulmonary vasculature.

Group 4: Chronic Thromboembolic Pulmonary Hypertension (CTEPH):

- **Etiology:** Caused by unresolved thromboemboli in the pulmonary arteries despite adequate anticoagulants.
- **Pathophysiology:** Chronic thromboembolic obstruction leads to increased pulmonary vascular resistance.

Group 5: Pulmonary Hypertension with Unclear Multifactorial Mechanisms:

- **Etiology:** Includes various conditions such as hematologic disorders, systemic disorders, metabolic disorders, and others.
- **Pathophysiology:** The mechanisms leading to pulmonary hypertension in this group are often complex and multifactorial.

Common pathophysiological features in PH include endothelial dysfunction, smooth muscle cell proliferation, inflammation, and fibrosis within the pulmonary vasculature.

Understanding the specific etiology and pathophysiology is crucial for proper diagnosis and management of pulmonary hypertension. Treatment approaches will **vary greatly** based on the underlying cause and the group to which the PH belongs.

DIAGNOSTIC CHALLENGES

The diagnosis of pulmonary hypertension (PH) in Canada currently can pose several challenges. Some of the key challenges include:

- **Healthcare System Challenges:** Factors such as long wait times for diagnostic tests, limited resources, and competing healthcare priorities can contribute to delays in the diagnostic process. This is particularly relevant in the context of echocardiograms (which are the critical screening tool of choice for PH) in some parts of the country, diagnostic imaging, right heart catheterization, and other specialized tests required for PH diagnosis.
- **Non-Specific Symptoms:** The symptoms of PH, such as shortness of breath, fatigue, and chest pain, can be non-specific and overlap with symptoms of other cardiovascular or respiratory conditions. This can contribute to delays in diagnosis and treatment.
- **Limited Awareness and Education:** Awareness among healthcare professionals and the general public about PH may be limited. As a result, healthcare providers might not always consider PH in the differential diagnosis, and patients may not seek medical attention promptly for symptoms.
- **Access to Specialized Centers:** Diagnosing and managing PH often require specialized expertise and diagnostic tools. Access to specialized PH centers, where comprehensive assessments and tests can be performed, may be limited in certain regions of Canada like Prince Edward Island or the North (where there is no specialized center). This can lead to delayed diagnosis or inadequate evaluation.
- **Variability in Diagnostic Practices:** There may be variability in the diagnostic practices and guidelines followed by different healthcare providers or institutions. Consistency in the approach to diagnosis and the use of standardized criteria are crucial for accurate and timely identification of PH.
- **Underdiagnosis in Specific Patient Groups:** PH can be underdiagnosed in certain patient populations, such as those with connective tissue diseases or chronic lung diseases. Recognizing and diagnosing PH in these groups may require a high level of suspicion and expertise.
- **Remote and Indigenous Communities:** Access to healthcare services, including specialized diagnostic facilities, can be challenging in remote and Indigenous communities. This may result in delayed diagnosis and limited access to appropriate treatments.

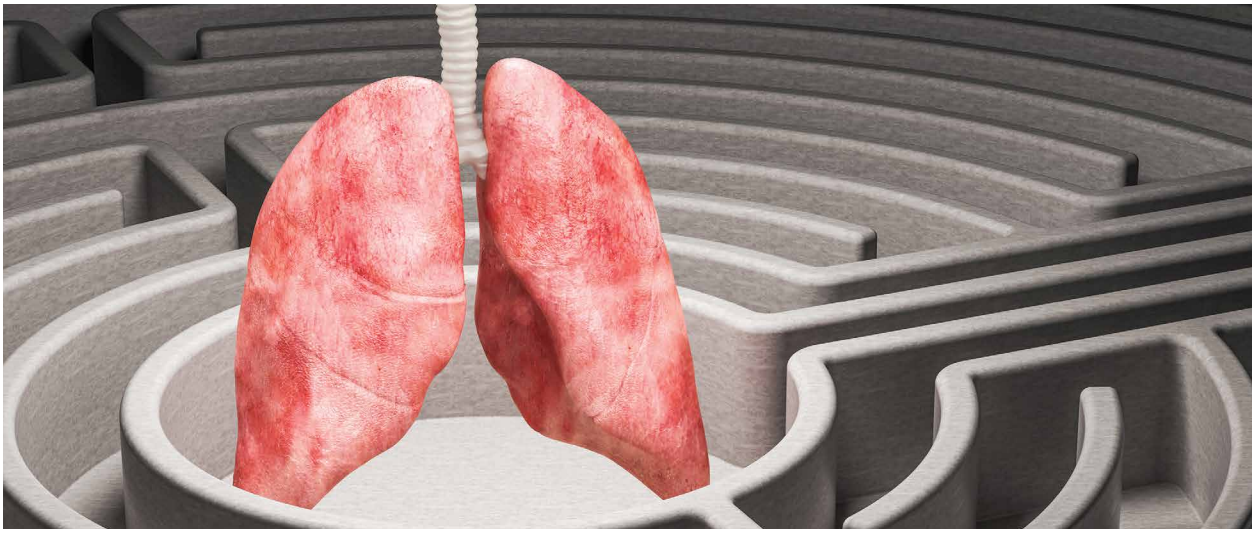
Addressing these challenges requires a multi-faceted approach, including increased awareness, education for healthcare professionals, improved access to specialized centers, and the development of standardized diagnostic pathways. **Collaboration among primary care providers, specialists, and patient advocacy groups is essential to enhance the diagnosis and management of pulmonary hypertension in Canada[5].**

I was in a mid-sized city recently in Quebec, a good city, good hospital. They said they could wait a year for an echo. Unless, the doctor specifically calls the ECHO lab and says, "I'm really worried about PH, speed it up". Otherwise, it's a year. Now for my patients to lose a year is catastrophic.

-David Langleben, Cardiologist

For our patients in the northern parts of Canada, it's very difficult for them to come. Some of our patients are very sick. The thought of having to drive eight or 10 hours to come and see us, it's just too hard for them physically. Alternatively, getting on a plane is also not easy for them, particularly if they're on oxygen. There's certainly a fair bit of reluctance to come. It's definitely proportional to the level of disability or illness of the patient. When patients are feeling reasonably well and they have to come twice a year to see us, we try to facilitate those visits during the favorable driving seasons, like spring and fall. But, when patients are not well and they have limited mobility, comorbidities, and are frail, it is very hard to get them to come.

-Lisa Mielniczuk, Cardiologist



DIAGNOSTIC TESTS USED FOR PH

Diagnosing pulmonary hypertension (PH) typically involves a combination of clinical evaluation and specific tests. Here's a brief summary of the key diagnostic tests patients with suspected PH may expect to undergo[6]:

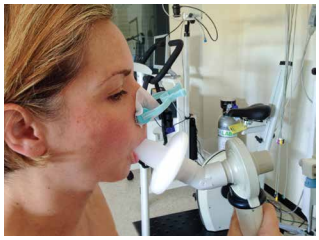
1. **Echocardiogram (Echo):** Sound waves are used to create pictures of the heart, lungs and chest. Provides a non-invasive assessment of cardiac structure and function, helping to assess the presence of PH and estimate pulmonary artery pressure.

2. **Electrocardiogram (ECG or EKG):** Records the heart's electrical activity, identifying signs of right heart strain or hypertrophy, which can indicate PH.

3. **Chest X-ray:** Helps visualize heart and lung changes, such as enlarged pulmonary arteries and right ventricular hypertrophy indicative of PH.

4. **Ventilation/perfusion (V/Q) scan:** A radioactive tracer is given through a vein (IV). The tracer shows blood flow. You also may breathe in a tracer that shows airflow to the lungs. A mismatch in blood flow and airflow can suggest that blood clots are causing PH.

5. **Computerized tomography (CT) scan:** A CT scan, can show the size of the heart and any enlargement or blockages in the pulmonary arteries. It can help diagnose lung diseases that might lead to PH such as COPD or pulmonary fibrosis.



6. Pulmonary Function Tests (PFTs): Measures lung function by blowing into a special device that measures how much air the lungs can hold and how it flows in and out of the lungs. PFTs can detect underlying lung diseases that may contribute to PH.

7. Sleep Study: Measures brain activity, heart rate, blood pressure, oxygen levels and body movements during sleep. This test can help diagnose obstructive sleep apnea, which can cause PH or make it worse.

8. Blood Tests: Screen for conditions contributing to PH, like connective tissue diseases, liver disease, HIV, methamphetamine use or blood clotting disorders.

9. Right Heart Catheterization (RHC): An invasive procedure in which a physician places a thin, flexible tube called a catheter into a blood vessel, usually in the neck. The catheter is gently guided into the lower right heart chamber and the pulmonary artery. A doctor can then measure blood pressure in the main pulmonary arteries and the right ventricle. It is essential for a **definitive PAH diagnosis** and assessing its severity.

These tests, in combination with clinical assessment, help evaluate the heart, lungs, and circulation, aiding in PH diagnosis and assessment of its severity and potential causes.

COMPREHENSIVE MANAGEMENT

The management of PH in Canada involves a comprehensive and multidisciplinary approach. Pulmonary hypertension is a condition characterized by elevated blood pressure in the pulmonary arteries, which can lead to various symptoms and complications. The management typically includes medical therapies, lifestyle modifications, and supportive care. Here's an overview:

1. Medical Therapies:

Medications are often prescribed to manage symptoms and slow the progression of certain types of PH. Optimizing treatment for underlying heart and lung diseases that is contributing to PH is very important. For PAH; vasodilators, endothelin receptor antagonists, prostacyclin analogs, and phosphodiesterase inhibitor may be prescribed. In some cases, anticoagulant medications can be prescribed to reduce the risk of blood clots. CTEPH can be treated with Pulmonary thromboendarterectomy (PEA) surgery and balloon pulmonary angioplasty (BPA). An sGC (soluble guanylate cyclase) stimulator can be prescribed for inoperable CTEPH.

2. Lifestyle Modifications:

- Exercise and Physical Activity: While exercise must be approached cautiously, supervised and tailored exercise programs can be beneficial for some individuals with PH.
- Dietary Changes: A heart-healthy diet is often recommended to manage overall cardiovascular health.
- Reduce Alcohol and Smoking Cessation: These substances can exacerbate the condition by causing additional strain on the heart and lungs, worsening symptoms, and potentially interfering with the medications used to manage PH.
- Avoidance of Triggers: Patients may be advised to avoid situations or substances that can exacerbate symptoms, such as air travel, high altitudes or certain medications.
- Vaccinations: Ensuring vaccinations are up to date is important to help prevent acute infectious illnesses that can exacerbate PH.
- Pregnancy: For women already diagnosed with PH, pregnancy poses significant dangers. PH tends to deteriorate during pregnancy, frequently resulting in maternal mortality. Thus, it is strongly advised that women with PH refrain from pregnancy and use effective contraception methods.
- Stress and Mental Health Management: Improves patients' quality of life, helps with adherence to treatment plans, and makes it easier to deal with the daily challenges of living with this condition.

3. Supportive Care:

- Oxygen Therapy: In cases of low oxygen levels, supplemental oxygen may be prescribed to improve oxygenation.
- CPAP: CPAP machines help to keep the airways open during sleep ensuring adequate oxygenation, reducing the frequency of apnea episodes, improving sleep quality, and alleviating symptoms associated with obstructive sleep apnea which can cause and/or make PH worse.

- Symptom Management: Medications may be used to manage specific symptoms like diuretics for fluid retention.
- Regular Monitoring: Patients with PH often require regular monitoring of their condition, including imaging studies, pulmonary function tests, and blood tests.

4. Specialized Care Centers:

Treatment and management of PH **may** be coordinated through specialized PH centers, whereas the treatment and management of PAH and CTEPH **must** be coordinated through specialized centers. These centers have expertise in the diagnosis and management of this complex condition.

- Access to experimental therapies and clinical trials may only be available in these specialized academic centers.

5. Transplantation:

- In severe cases where medical therapy is not sufficient, lung transplantation may be considered as a treatment option.

It's important to note that the specific treatment plan for an individual with PH is determined by the severity of the condition, the underlying cause, and the patient's overall health. Management is typically tailored to each patient's specific needs, and regular follow-up with healthcare providers is crucial to monitor the progression of the disease and adjust the treatment plan accordingly.

I think one of the biggest challenges is probably access to not only the diagnostic tests, but also, the test that we use to follow people so that we can monitor them effectively. Especially, when they live far away (for example, northern Canada). So, I think the equity of accessibility to tests is the biggest problem in Canada in terms of PH management.

Jason Weatherald, Respirologist

The Ministry of Health in Ontario is pretty strict about the way we are allowed to order drugs to treat PAH, for example selexipag has to be ordered third, it can't be ordered second. We're grateful that the companies may compassionately provide, but it can be frustrating at times that it is so difficult to access these important drugs.

Carolyn Doyle-Cox,
Advanced Practice Nurse

REFERRALS

In Canada, patients with PAH are referred to specialized centers by healthcare professionals, often by specialists such as cardiologists or respirologists, while some patients with PH may be referred by their primary care physician.

These specialized centers, known as Pulmonary Hypertension clinics or centers, have expertise in the diagnosis and management of PH.

Referrals to specialized PH centers can come from various healthcare providers involved in the care of the patient, including:

1. Cardiologists: Since PH involves the cardiovascular system, cardiologists are often involved in the diagnosis and management. Cardiologists may refer patients to specialized PH centers for further evaluation and treatment.
2. Respirologists/Pulmonologists: Given the impact of PH on the respiratory system, respirologists or pulmonologists may be involved in the care of these patients. They may refer individuals with suspected or confirmed PH to specialized centers for comprehensive care.
3. Rheumatologists: In cases where PH is associated with connective tissue diseases, rheumatologists may be involved in the referral process.
4. Primary Care Physicians: Nurse practitioners and family physicians may also play a role in referring patients to specialized centers if they suspect or diagnose PH.
5. Surgeons: Playing a role in diagnosing and treating CTEPH, surgeons can perform a pulmonary endarterectomy (PEA). This surgical procedure is aimed at removing chronic blood clots from the pulmonary arteries. It is specialized surgery, typically performed by cardiothoracic surgeons with expertise in treating CTEPH. Balloon Pulmonary Angioplasty (BPA) is an investigational procedure to treat CTEPH that is done by an interventional radiologist to dilate pulmonary arteries that are obstructed by the chronic scar tissue.
6. Other Specialists: Depending on the underlying cause of PH, other specialists such as hematologists, hepatologists, or infectious disease specialists (HIV) may be involved and contribute to the referral process.

Referral to a specialized PH center ensures that patients receive care from a multidisciplinary team with expertise in the specific needs of individuals with Pulmonary Hypertension. These centers often have access to advanced diagnostic tools, specialized treatments, ongoing monitoring to optimize patient outcomes, and conduct research studies to advance our understanding of and treatment options for PH.

The specific process for referral may vary by province or territory within Canada, making the process difficult.

Find a PH Center – Pulmonary Hypertension Association of Canada (phacanada.ca)

TREATMENTS

Medical treatments for PAH and CTEPH typically aim to alleviate symptoms, improve quality of life, slow the progression of the condition, and improve survival. However, there is no cure to halt or reverse the progression of the disease. The specific approach to treatment may vary depending on the underlying cause of PH. Regular monitoring and adjustments to the treatment plan may be necessary to optimize its effectiveness. Treatments can be administered orally, by sub-cutaneous injection or by IV infusion. They can be complex to administer and require significant expertise from the health care team.

The next couple of pages list the current treatments for PAH and CTEPH in Canada (by family/type), which are usually used in combination to treat most PAH patients[7].

PHARMACOLOGICAL TREATMENTS

- **Endothelin receptor antagonists (ERAS):** These drugs block the effects of endothelin, a substance that constricts blood vessels and promotes cell growth. By blocking endothelin receptors, ERAs help dilate blood vessels and reduce pulmonary vascular resistance, ultimately improving blood flow in the lungs. **Used in the treatment of PH group 1.**

- **Phosphodiesterase inhibitors (PDE-5 inhibitors):** PDE-5 inhibitors work by blocking the enzyme phosphodiesterase-5, which regulates the breakdown of cyclic guanosine monophosphate (cGMP). Increased levels of cGMP lead to relaxation of smooth muscle cells in the pulmonary arteries, resulting in vasodilation and improved blood flow. **Used in the treatment of PH group 1.**

- **Soluble guanylate cyclase stimulators (sGC stimulators):** These medications stimulate the enzyme soluble guanylate cyclase, which increases the production of cGMP. Like PDE-5 inhibitors, sGC stimulators promote vasodilation in the pulmonary arteries, reducing pulmonary vascular resistance and improving exercise capacity. **Used in the treatment of PH groups 1 and 4.**

- **IP receptor agonists:** IP receptor agonists activate the prostacyclin receptor (IP receptor), leading to vasodilation, inhibition of platelet aggregation, and suppression of smooth muscle cell proliferation in the pulmonary arteries. This helps to reduce pulmonary artery pressure and improve symptoms in patients with PH. **Used in the treatment of PH group 1.**

- **Prostacyclin analogues:** Prostacyclin analogues mimic the effects of prostacyclin, a naturally occurring vasodilator and inhibitor of platelet aggregation. By increasing prostacyclin levels, these drugs help dilate pulmonary blood vessels, reduce pulmonary artery pressure, and improve blood flow. **Used in the treatment of PH group 1.**

TREATMENTS FOR CTEPH (I.E. GROUP 4 PH)

SURGICAL TREATMENTS

- **Surgical treatment:** Surgical treatment, such as pulmonary endarterectomy, is a specialized procedure used to treat chronic thromboembolic pulmonary hypertension (CTEPH). It involves removing fibrosis from inside the pulmonary arteries, relieving obstruction and improving pulmonary blood flow.

- **Ballon Pulmonary angioplasty (BPA):** During BPA, a catheter is inserted into the blood vessels and a balloon is inflated to open up the vessel and increase blood flow throughout the lungs. BPA can be repeated multiple times to target different areas of the pulmonary arteries. It is an investigational procedure and the only treatment center for BPA in Canada is the Toronto CTEPH Program.

MEDICAL TREATMENTS

- **Soluble guanylate cyclase stimulators (sGC stimulators):** These medications stimulate the enzyme soluble guanylate cyclase, which increases the production of cGMP. Like PDE-5 inhibitors, sGC stimulators promote vasodilation in the pulmonary arteries, reducing pulmonary vascular resistance and improving exercise capacity.

Some provinces require PH patients to be followed by specialists/center to access insurance coverage for PH medications. Access to certain medications can be limited despite approval and depending on the province and insurance coverage. This means that people in some parts of the country do not have equitable access to treatments that they need. Shining a light on the disparities in access, availability and affordability of PH medications across Canada is important[1].

So, when I started, there was just not that much. Now we have more. And a new drug is coming that's even going to be a totally different pathway. So, there's a lot of growth in this field.

Carolyn Doyle-Cox,
Advanced Practice Nurse

I think the real goal is to find new therapies that truly do reverse the process or prevent it completely from progressing. PAH is negatively affecting the lives of people, who often are relatively young.

Paul Hernandez,
Respirologist

COMPREHENSIVE NEEDS ASSESSMENT

RESPIPLUS conducted a comprehensive needs assessment led by a Scientific Committee to identify areas requiring improvement, such as timely diagnosis, access to specialized care, and education. Based on the results, healthcare systems can work towards bridging these gaps and enhancing the quality of care provided to PAH patients across the country.

RESPIPLUS formed a **Scientific Committee of PH healthcare experts representing multiple Canadian organizations** and interviewed them to create our needs assessment survey, which was distributed to healthcare professionals across the country with the ultimate goal of producing this report.

In the preliminary stages, our Scientific Committee defined specific objectives for the report, ensuring a practical and attainable focus. This process involved aligning our goals with the insights sought from a diverse group of health experts, emphasizing a grounded approach to address real-world issues.

Our team was able to bring together a diverse panel of PH experts, engaging them in virtual panel meetings, interviews, and survey creation. This collaborative effort allowed us to gather valuable insights, document responses, and identify common themes.

Subsequent phases involved a comprehensive analysis of the survey data, where patterns and key takeaways emerged. Armed with these insights, the Scientific Committee crafted a practical and applicable paper, not just meeting but surpassing our initial objectives. Rigorous review by the PH experts refined our paper, leading to its finalization and publication, marking a tangible step forward in advancing practical knowledge within the realm of PH.

SURVEY KEY FINDINGS

The survey was open between January 12 and January 27, 2024, and was available in both English and French. It was distributed via email and social media channels to RESPIPLUS' member list, reaching over 5,000 healthcare professionals in Canada. The survey was additionally shared with CTS and FPAGC members as well as the various networks of our scientific panel.

There was a total of 147 respondents, 13 were excluded because they were not a practicing HCP in Canada.

Identified Care Gaps

- Lack of exposure and education about PH (in medical school and other training)
- Early diagnosis and targeted population screening
- Long wait times for diagnostic tests (especially echocardiograms) and referrals
- Inadequate access to specialized PH centers (often due to distance)
- Timely referral and access to therapies (PH medications)
- Unequal access for all patients to PH medications
- Insufficient patient education and support

To access the detailed survey results please see **Appendix I.**

ONGOING CHALLENGES IDENTIFIED BY HEALTHCARE PROFESSIONALS

A number of gaps or challenges remain in properly addressing the significant issues many patients face as a result of PH being poorly recognized and managed in the Canadian health care system.

Main categories of ongoing challenges according to the experts we interviewed include the following:

- Diagnostic delays and under recognition
- Long wait times for echocardiograms in many areas of Canada
- Inadequate disease awareness, education, and research
- Limited access to specialized care centers
- Fragmented care coordination and follow-up
- Barriers accessing treatment and therapies

For a more detailed list of the ongoing challenges in each of the above categories identified by our experts and respondents of our survey, please see **Appendix III.**

FIRST-HAND EXPERIENCE

What do we really need to help diagnose and treat PH patients according to the Canadian physicians, specialists, and healthcare professionals we interviewed:

The biggest need is more effective medications. At the end of the day, I'm a doctor and my job is to make people better. And while we have medications that buy people time, until recently we've had nothing that really deals with the underlying disease process. A critical issue to highlight is the significant delays in the approval process for new medications, compounded by the inefficient approval process at CADTH (Canadian Agency for Drugs and Technologies in Health). Many provinces lack access to all federally approved medications. This situation forces caregivers to make treatment decisions for a rare and fatal disease based on payer constraints rather than clinical considerations. With the potential introduction of new biologics to the market, this issue is likely to worsen, exacerbating the challenges faced by patients and healthcare providers.

-David Langleben, Cardiologist

We need more research. More understanding of the person, of the sort of the precision aspects of treating patients. Right now, it's just all about how many drugs we give patients. Do we give them one drug, two drugs, three drugs, four drugs now maybe? And I do wonder if we understood the disease a little bit better, we could be more precise. Based on certain characteristics or certain testing or biology. this patient should have this combination and they're going to be more likely to respond because unfortunately not every patient responds the same way to the therapies and we don't really know why that is yet.

-Lisa Mielniczuk, Cardiologist

I think we need more patient support groups. Patients are really good advocates for themselves if we allow them. With a support group, you can have education, like for example bringing in speakers several times a year. The patients can learn from each other, get involved in fundraising, and in going to talk to their elected leaders about issues like unequal access to therapies. PH patients are the most knowledgeable population of patients that I have ever taking care of.

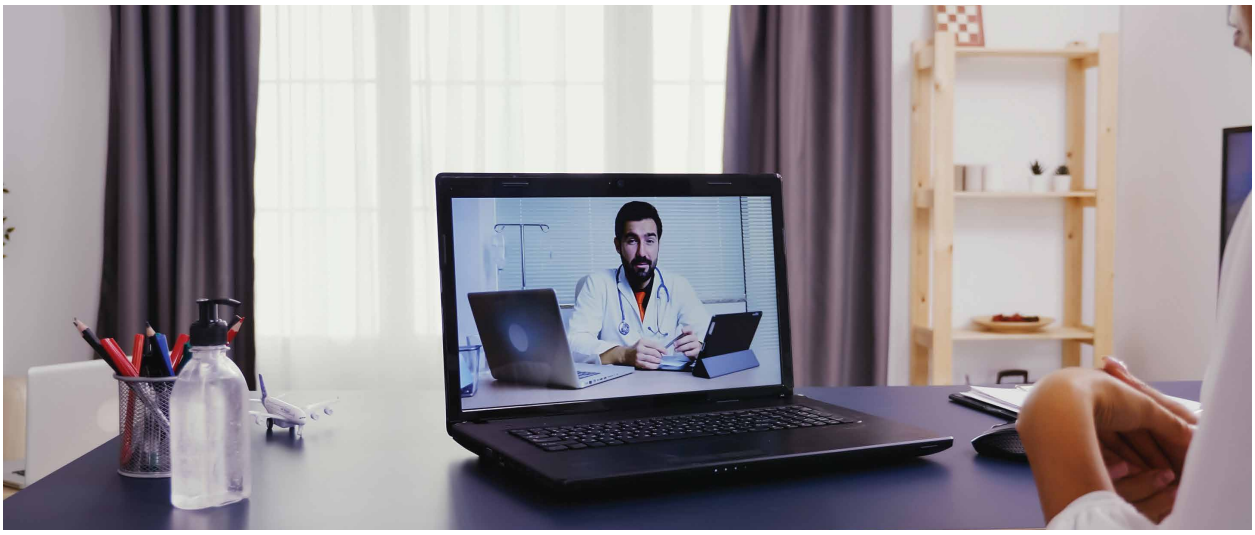
-Carolyn Doyle-Cox, Advanced Practice Nurse

RECOMMENDATIONS AND ACTION PLAN

I. IMPROVING CARE FOR PH
II. EDUCATIONAL INITIATIVES
III. THE ROAD AHEAD
IV. ACTION PLAN

IMPROVING CARE FOR PH

The following section outlines recommendations and an action plan aimed at enhancing PH care within Canada. Drawing upon insights gathered from our PH experts, survey analysis and collaborative efforts, the proposed strategies seek to address critical areas of improvement, streamline care pathways, and enhance PH patient outcomes.



1. Clinical Guidelines and Protocols:

- The committee found that there is a need to disseminate evidence-based clinical guidelines, such as those of the CTS and other medical societies, and protocols for the timely diagnosis, management, and referral of patients with suspected PH.
- However, a continuous educational intervention is needed to ensure that these guidelines are easily accessible and regularly updated for healthcare professionals across different specialties.

2. Streamlined Referral Pathways:

- There is a need for increased awareness of PH in specific groups such as primary care providers, as they need to regularly screen for PH in certain high-risk patient populations.
- Provincial health authorities could establish streamlined referral pathways for patients suspected of having PH to ensure timely access to specialized PH clinics. PHA Canada has done great work in highlighting excellent PH centers across the country.
- This process could be facilitated by electronic referral systems and standardized referral forms to facilitate communication between primary care providers and PH specialists.

3. Telemedicine and Virtual Consultations:

- An effort will need to be put in place to expand telemedicine services and virtual consultations to improve access to PH specialists for patients in remote or underserved areas.
- As well, we need to provide training and support for healthcare professionals to effectively conduct virtual consultations and manage PH patients remotely. CTS could help promote research, education and guidelines related to telemedicine in respiratory care.

4. Multidisciplinary Care Teams:

- The committee would highly encourage the establishment of multidisciplinary PH care teams comprising of specialists from cardiology, respirology, rheumatology, nursing and other relevant disciplines across Canada.
- Foster collaboration and communication among team members to ensure comprehensive and coordinated care for PH patients. PHA Canada is dedicated to supporting individuals with PH and can play a vital role in fostering collaboration among healthcare providers.

5. Data Collection

- Implement standardized data collection tools and registries like the Canadian Pulmonary Hypertension Registry (CPHR) to monitor PH epidemiology, outcomes, and quality of care across Canada.

EDUCATIONAL INITIATIVES

In this section we want to highlight the essential educational initiatives required to raise awareness, understanding, and management of PH. Recognizing the pivotal role of education plays in early detection, effective treatment, and supportive care, the following recommendations aim to empower healthcare professionals, patients, caregivers, and the broader community with important knowledge and skills with regards to PH.

Training and Continuing Medical Education (CME)

There is a continuous need for **CMEs, webinars opportunities and training programs** targeting all healthcare professionals to improve their knowledge and skills in diagnosing and managing PH. The organizations preparing this report have great expertise in the development and dissemination of such programs via online websites, webinars, conferences, etc. To increase the impact, we will need to incorporate PH education into medical schools, residency curricula and other healthcare professional education programs to ensure that future generations of healthcare professionals are well-equipped to identify and treat PH. Collaborations with **Universities** are needed.

Education and Awareness Campaigns

There is a need to launch national and regional campaigns to raise awareness about PH among healthcare professionals, patients, and the general public. RESPIPLUS has a vast community of +25,000 members across Canada and the world and would be a great organization to support awareness across a wider healthcare community. Many other organizations, such as medical associations, patient advocacy groups, and government health agencies can continue to disseminate information about PH symptoms, risk factors, and the importance of early diagnosis and treatment.

Patient Empowerment and Support Groups

Members of our committee saw a need to launch national and regional campaigns to raise awareness about PH among healthcare professionals, patients, and the general public. PHA Canada can play a leading role in patient awareness initiatives.
We should encourage collaboration with medical associations, patient advocacy groups, and government health agencies to disseminate information about PH symptoms, risk factors, and the importance of early diagnosis and treatment. CTS can help develop guidelines, educational materials, and training programs for healthcare professionals regarding PH recognition, diagnosis, and management. As well, patient support groups and organizations such as PHA Canada can allow patients to share their experiences and knowledge and help them advocate for themselves for better treatments and management at the government level. Established PH clinics across Canada can help start and encourage patient led support groups within their communities.

THE ROAD AHEAD

The Scientific Committee has discussed areas of improvement to enhance healthcare policies for PH treatment and management. We are listing here several changes and collaborations that can be considered.

Policy Enhancements:

- National PH Strategy: Develop a national strategy for PH that outlines standardized protocols for early detection, diagnosis, and management of the condition across all provinces and territories.
- Access to Specialized Care: Implement policies to ensure timely and equitable access to echocardiograms, specialized PH care centers and multidisciplinary teams equipped to provide comprehensive care and support to patients.
- Patient-Centered Care: Emphasize patient-centered care in healthcare policies by promoting shared decision-making, patient education, and empowerment throughout the treatment journey. In particular, PHA Canada has been promoting this commitment via their organization.
- Integration of Telemedicine: Facilitate the integration of telemedicine services into PH care delivery to improve access to specialized expertise, enhance monitoring, and support remote patient management.

Collaboration with Healthcare Providers and Systems:

- Interdisciplinary Care Teams: Encourage collaboration among healthcare providers, including respirologists, cardiologists, nurses, pharmacists, and allied health professionals, to establish interdisciplinary care teams dedicated to PH management. Multiple centers in Canada where PH care is provided use this approach and perhaps other clinics could learn from their expertise.
- Continuing Education and Training: Provide ongoing education and training opportunities for healthcare providers to enhance their knowledge and skills in PH diagnosis, treatment, and management. This can be done via accredited and/or non-accredited learning opportunities. CTS and RESPIPLUS are key partners with vast expertise in this area.

Partnerships with the Pharmaceutical Industry and Collaborators:

- Research and Development Funding: Collaborate with the pharmaceutical industry to support research and development efforts aimed at advancing the understanding of PH pathophysiology, identifying novel treatment targets, and developing innovative therapies.
- Patient Assistance Programs: Partner with pharmaceutical companies to establish patient assistance programs that provide increased financial assistance, medication access, and support services to patients with PH who may face barriers to treatment.

Clinical Trials and Data Sharing:

- Encourage collaboration among healthcare providers, research institutions, and pharmaceutical companies to conduct clinical trials, collect real-world data, and share insights to inform evidence-based practice and improve patient outcomes.
- Non-Profit and Professional Organizations: Partner with non-profit organizations and professional associations, such as patient advocacy groups (such as PHA Canada), medical societies (such as CTS and FPAGC), and research foundations, to leverage their expertise, resources, and networks in raising awareness, providing support, and advocating for policy changes related to PH.

We expect that by implementing these strategies and fostering meaningful collaborations across healthcare sectors, policymakers, healthcare professionals, industry stakeholders, and patient advocates can work together to address gaps in PH care delivery, improve patient outcomes, and ultimately enhance the quality of life for individuals living with pulmonary hypertension.

ACTION PLAN

In this comprehensive report, our primary aim is to elevate awareness of pulmonary hypertension (PH) in Canada while striving to enhance patient outcomes through tailored educational strategies. We propose leveraging PHA Canada's existing Leadership Council comprised of patient partners, PH medical experts, and researchers to spearhead initiatives aimed at reducing diagnostic delays and improving access to specialized care across the country.

By allocating resources to support targeted interventions, such as the establishment of additional multidisciplinary care teams and the integration of more telemedicine services, we can ensure equitable access to high-quality care for all Canadians affected by PH. Moreover, it is essential to establish mechanisms for ongoing evaluation and feedback to continuously refine and enhance PH care delivery.

Additionally, **we advocate for policy changes to prioritize PH as a national health concern,** ensuring that it receives the attention and resources necessary for effective management. Through these concerted efforts, we envision a future where PH is recognized, diagnosed, and managed promptly and effectively, ultimately improving the lives of patients and their families across Canada.

A variety of informational and educational tools should be considered including the following:

- Webinars and podcasts provided by experts
- In-person or virtual conferences
- Web-based continuing education programs

Survey data of Canadian HCPs showed support for receiving information on the following topics:

- How to maintain a healthy lifestyle with PH
- What to do if your symptoms get worse
- How to exercise and be physical active with PH
- Guidance on how to live positively
- Guidance on how to navigate the healthcare system to receive diagnosis

CONCLUSION

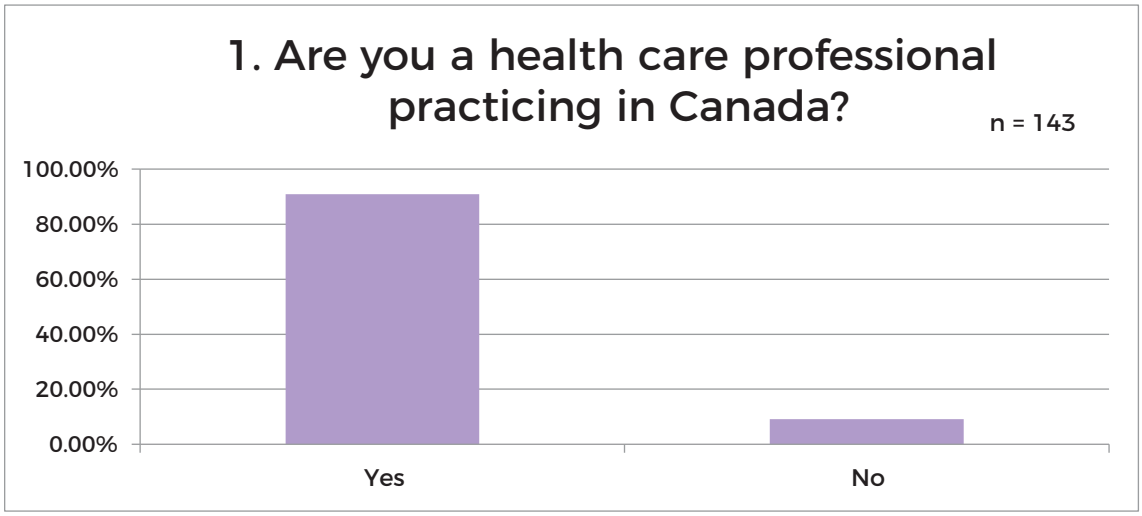
Pulmonary Hypertension (PH), characterized by elevated blood pressure in the arteries of the lungs, poses significant challenges to patients and healthcare systems worldwide. Despite advances in medical science, many patients still face considerable barriers to optimal care. The burden of PH extends beyond physiological symptoms, affecting mental health, social dynamics, and economic stability.

Improving the care of people living with PH patients demands a collaborative approach. Healthcare providers, policymakers, and industry partners must unite to drive meaningful change across the care continuum. By leveraging the collective expertise, resources, and influence, we can pioneer transformative solutions that empower people with PH and enhance their well-being.

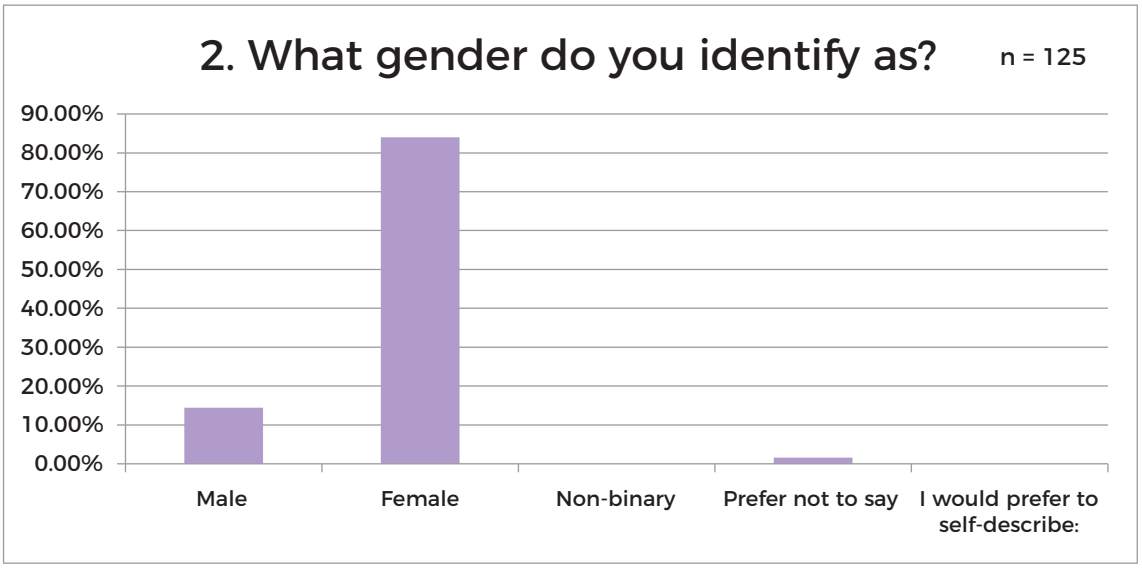
APPENDIX

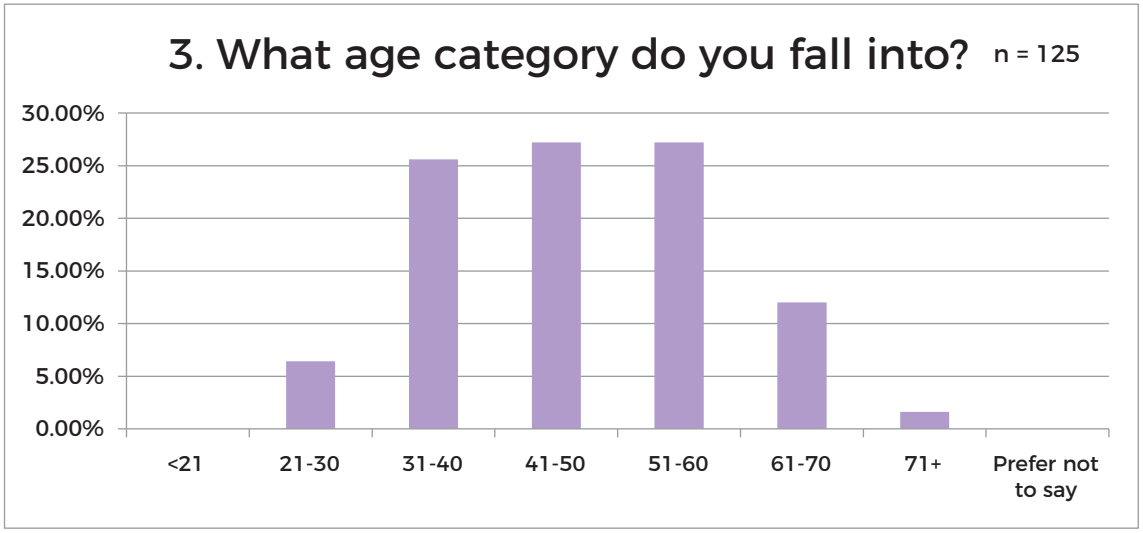
I. SURVEY RESULTS
II. TOOLS & RESOURCES
III. ONGOING CHALLENGES

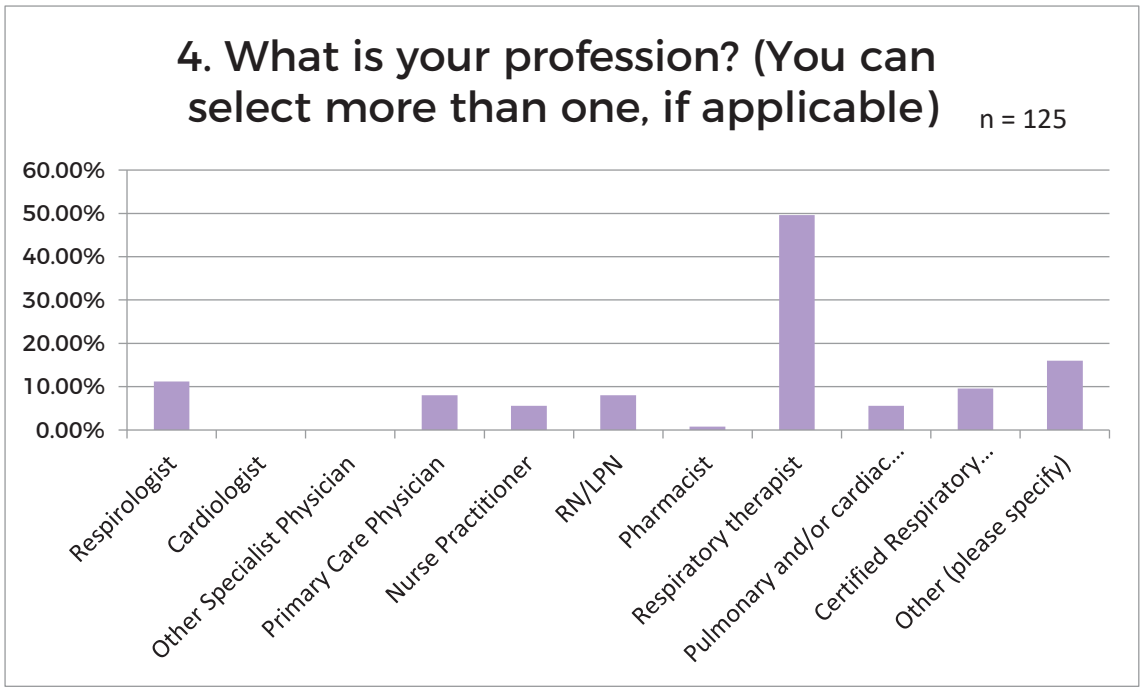
APPENDIX 1 – SURVEY RESULTS

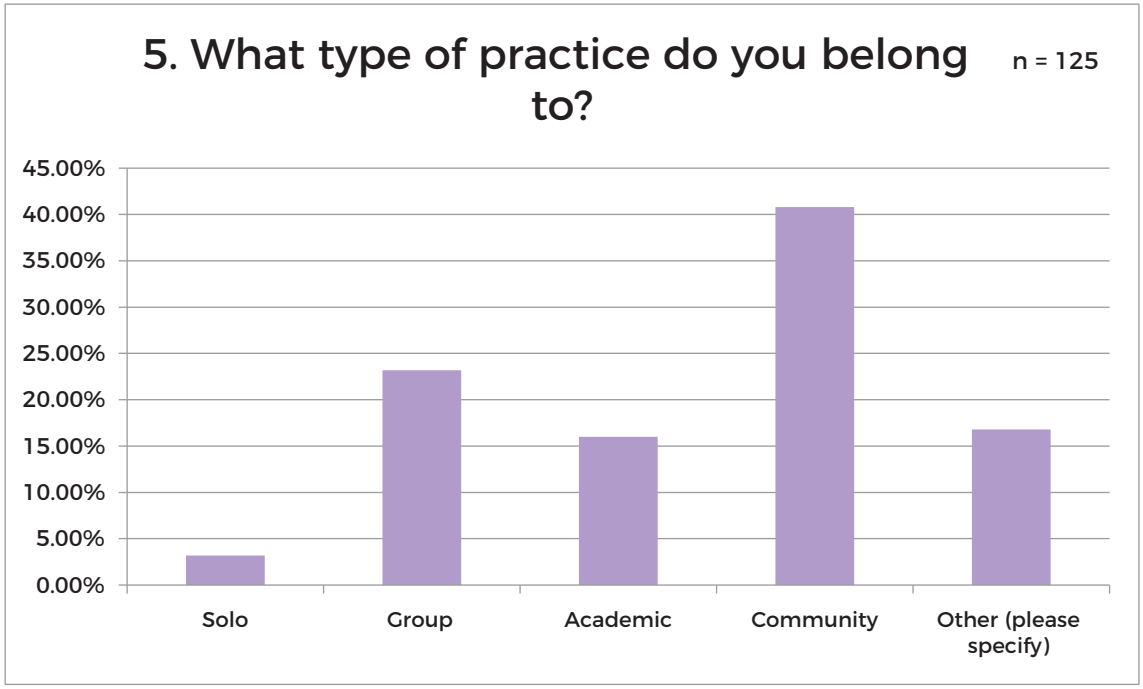


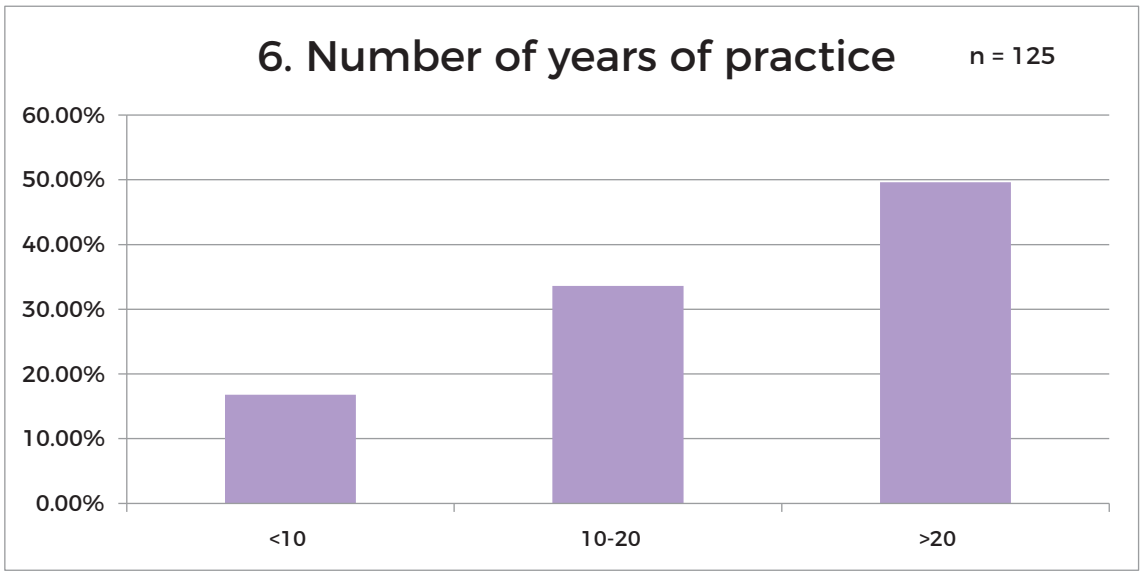
*13 excluded for not being a practicing HCP in Canada, 5 dropped out.

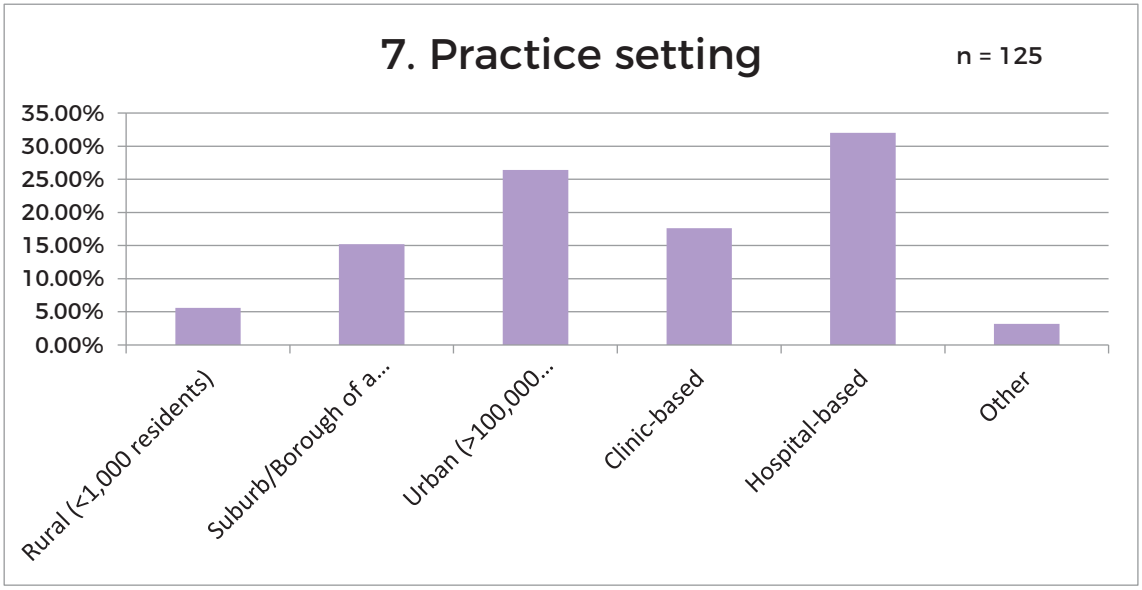


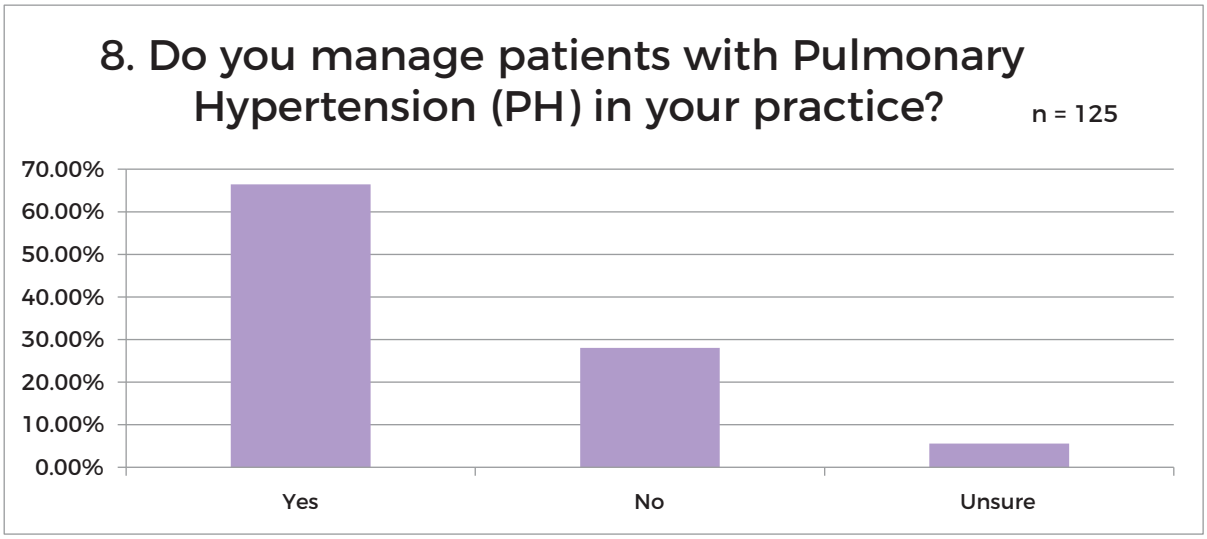


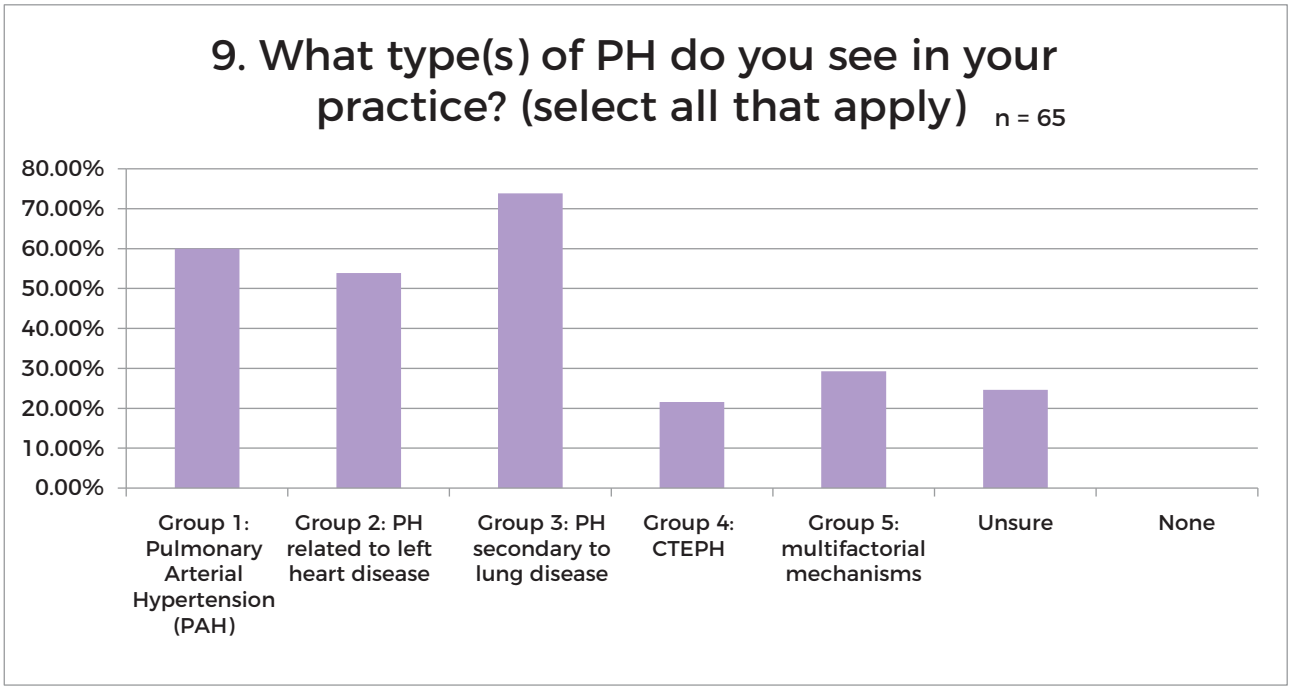




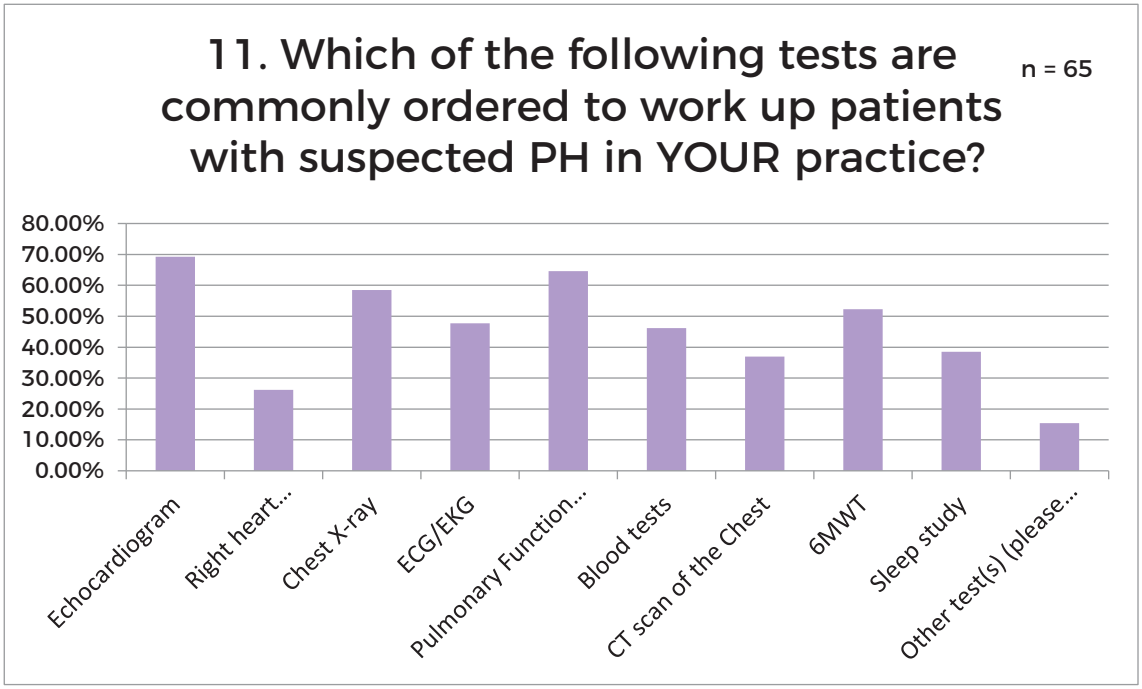






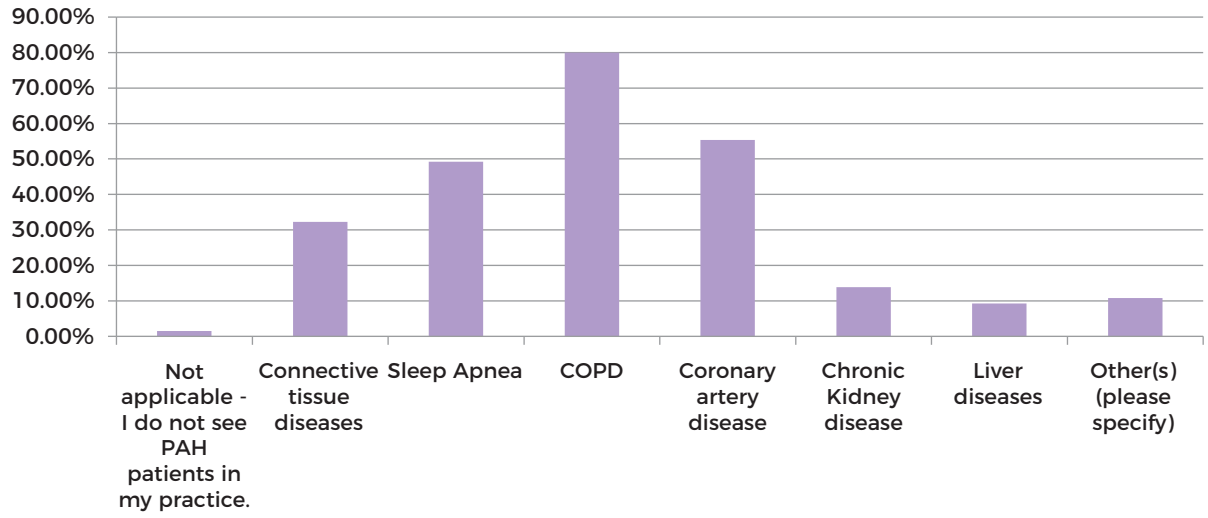


10. Approximately how many patients with PH, from any cause, do you see per month? (Enter a whole number) n = 41
AVG = 8



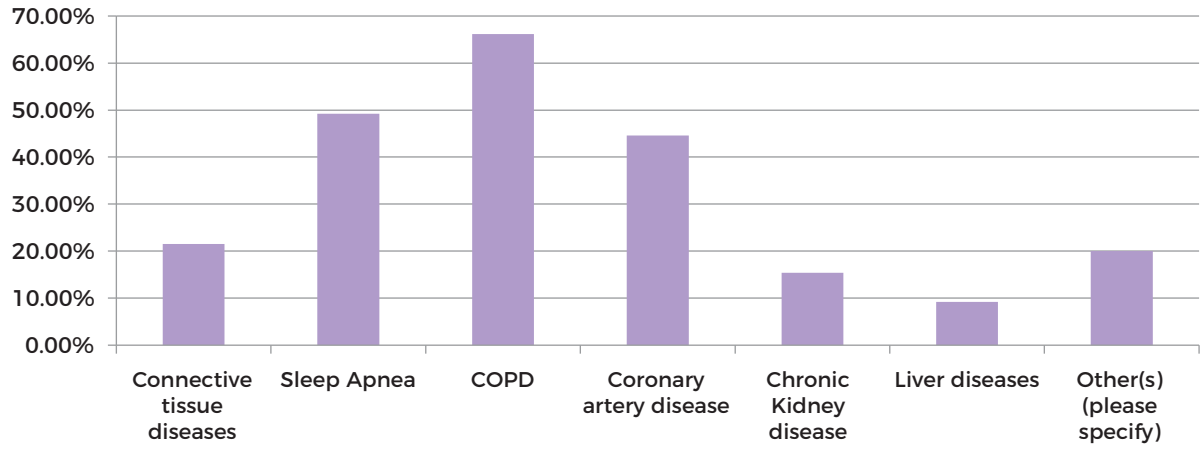
12. Which of the following are the most common comorbidities or conditions that you see in patients with confirmed PAH?

n = 65

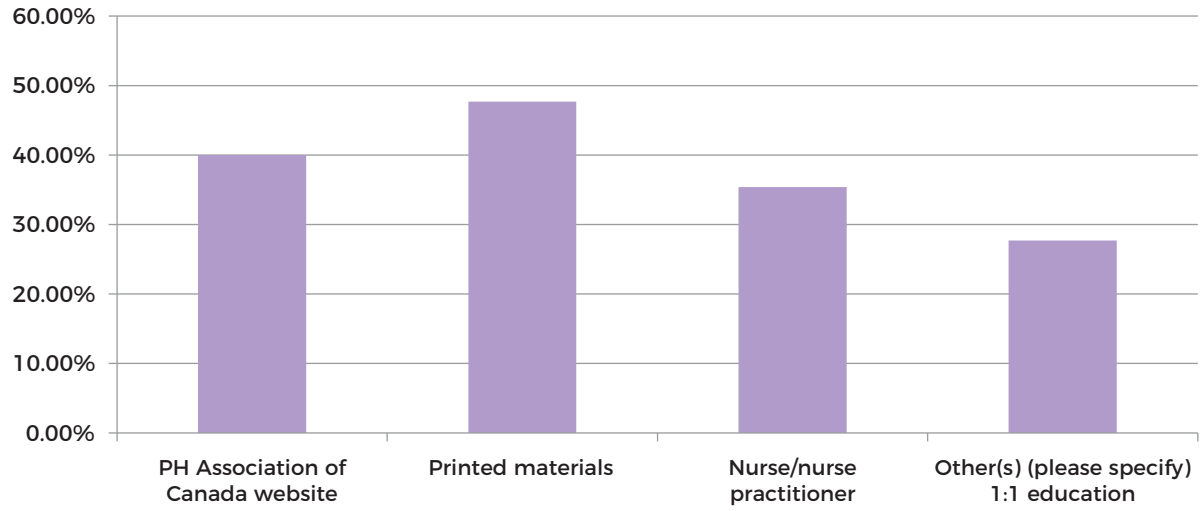


13. Which of the following are the most common comorbidities or conditions that you see in patients with non - PAH PH?

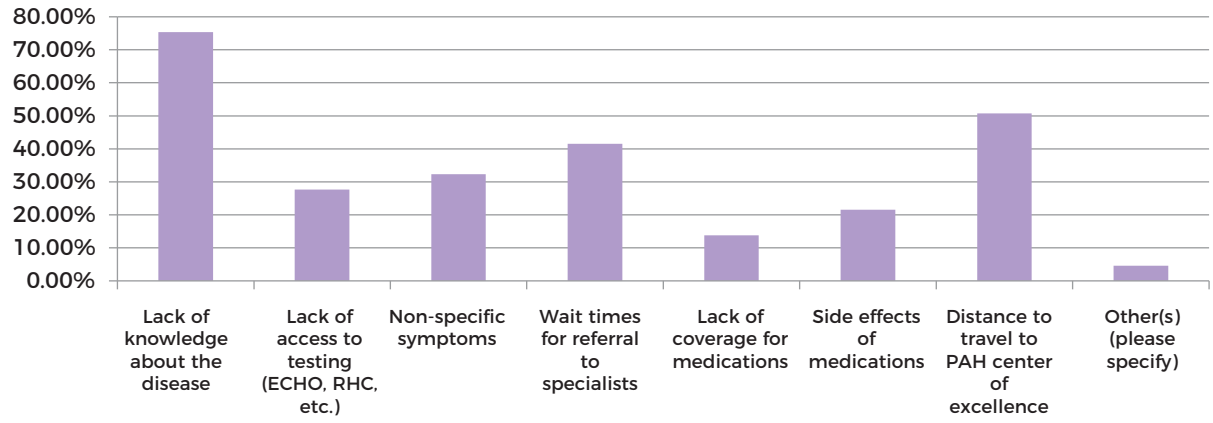
n = 65



14. What approach or resources do you use to educate and support those with PAH? n = 65

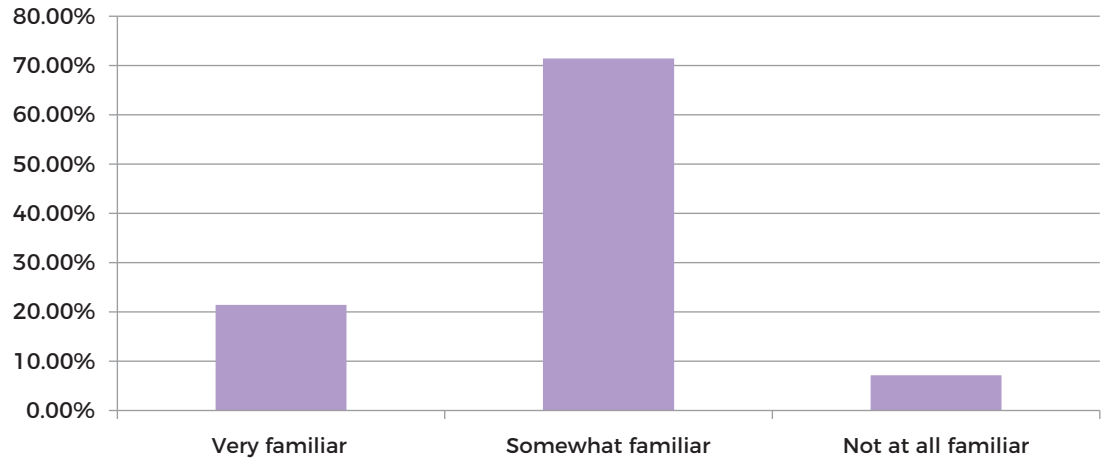


15. What are some of the most significant challenges you face when treating PAH, both in terms of diagnosis and management? n = 65



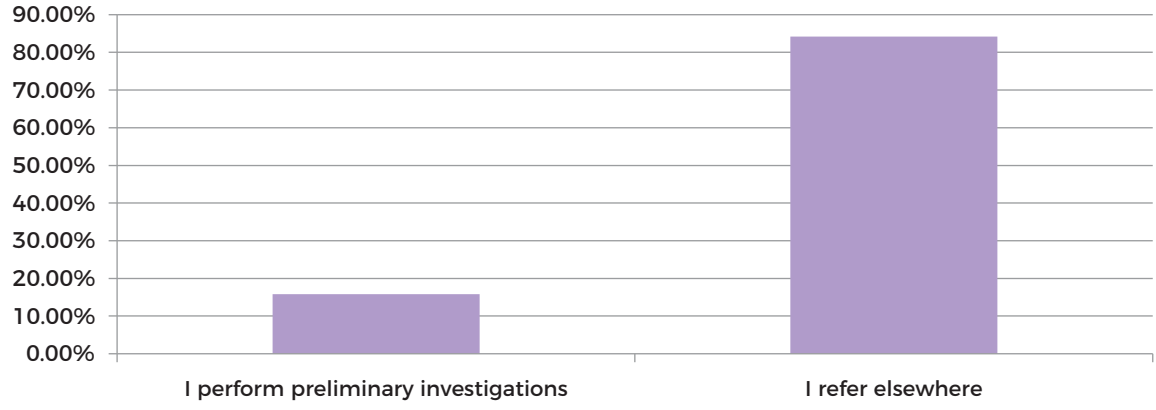
16. If you do not manage PH patients in your practice, are you still familiar with this condition?

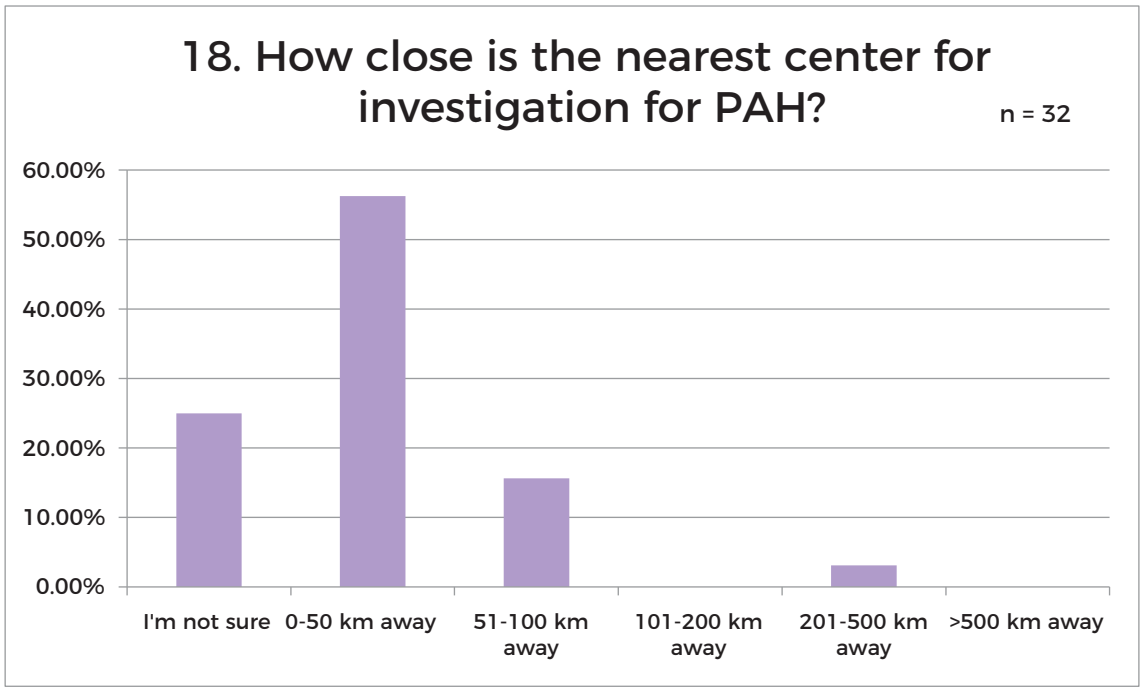
n = 42

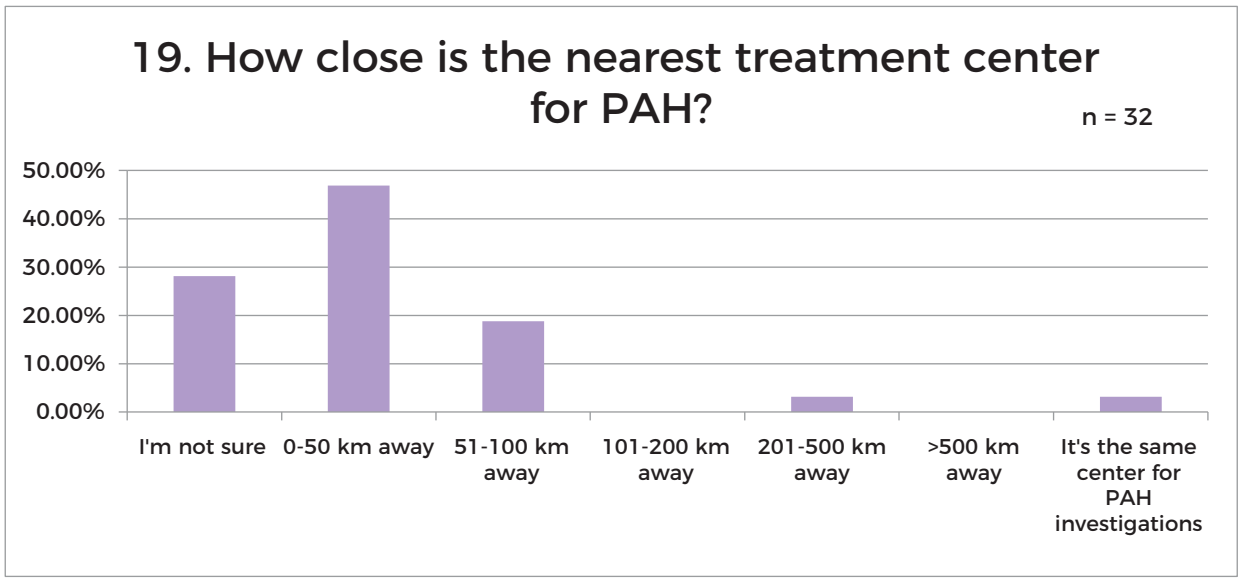


17. Talking more specifically about Pulmonary Arterial Hypertension (PAH), do you do investigations required for PAH, or do you refer elsewhere when suspected?

n = 38

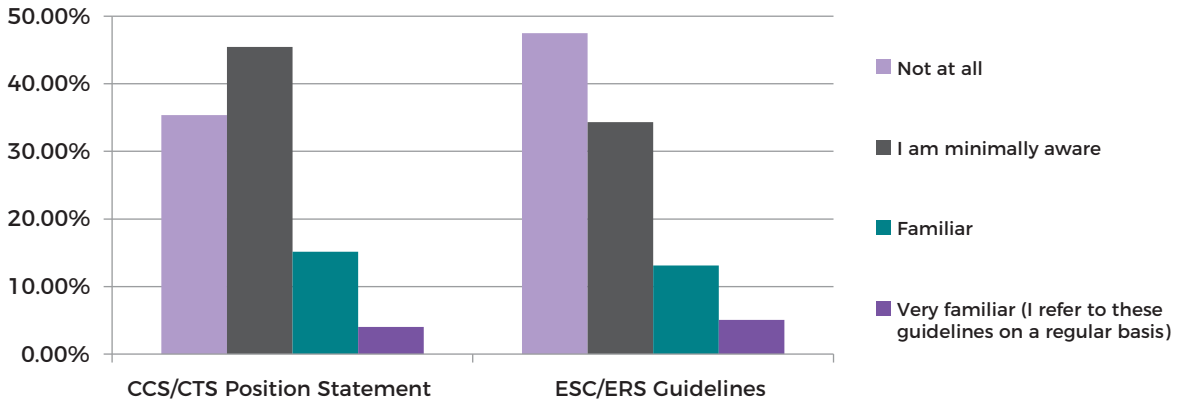






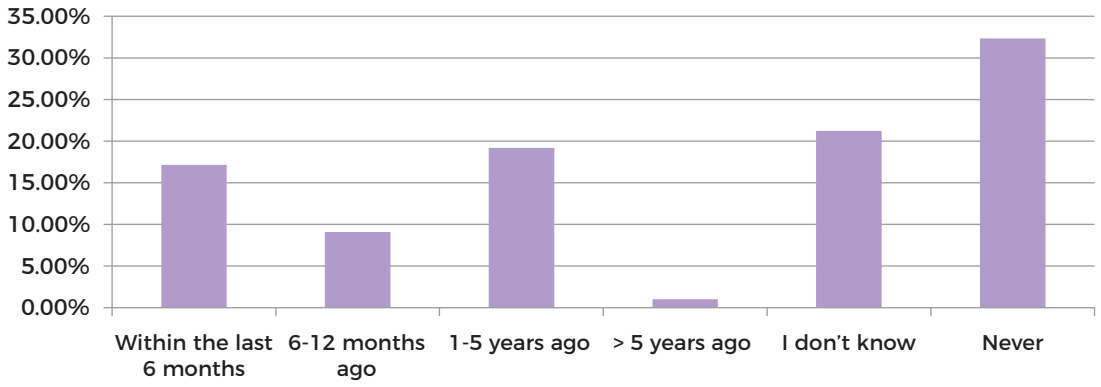
20. How familiar are you with the following guidelines and/or position statements for the management of PAH?

n = 99



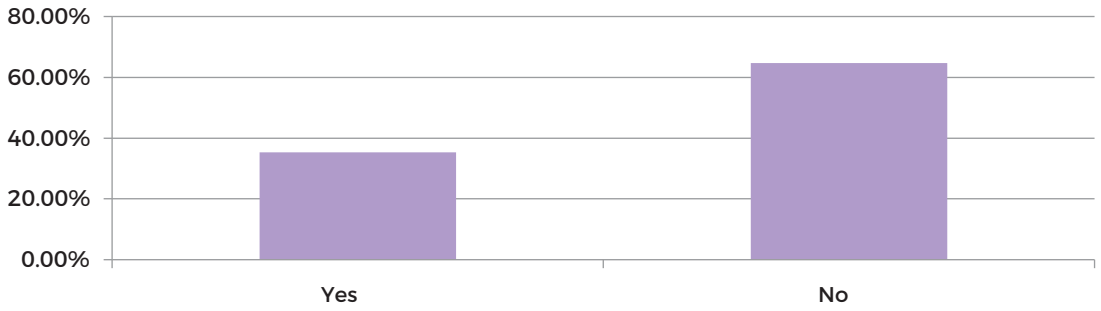
21. When was the last time you consulted either of the previously listed guidelines/position statement?

n = 99



22. Have you had training or education about PAH previously?

n = 99

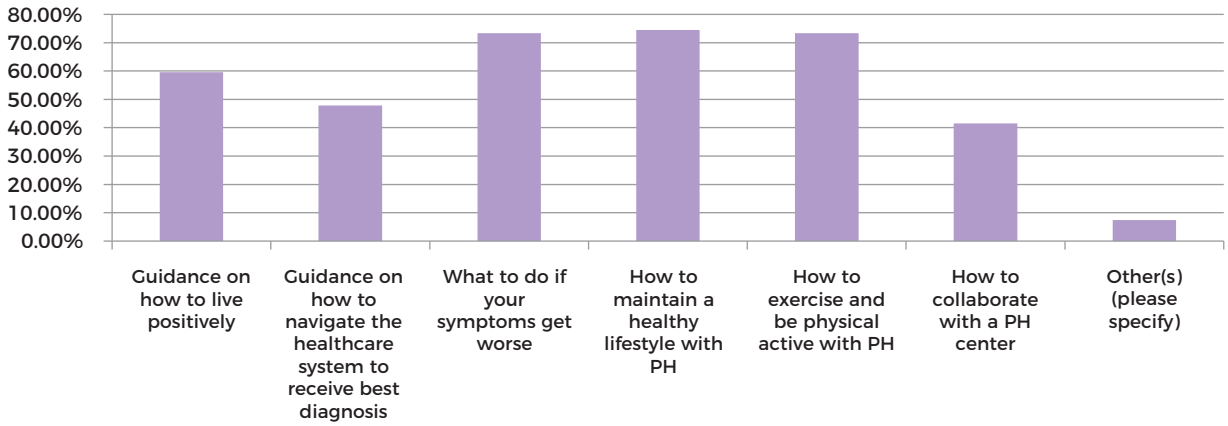


23. Where did you undergo this training/education?

n = 35

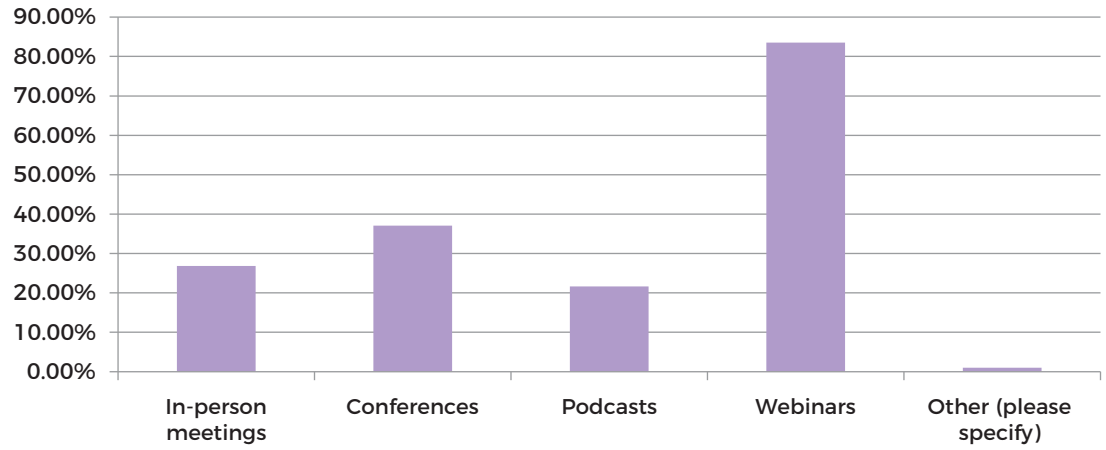
24. Which content topics would be most useful for you to learn and use with your PH patients?

n = 94



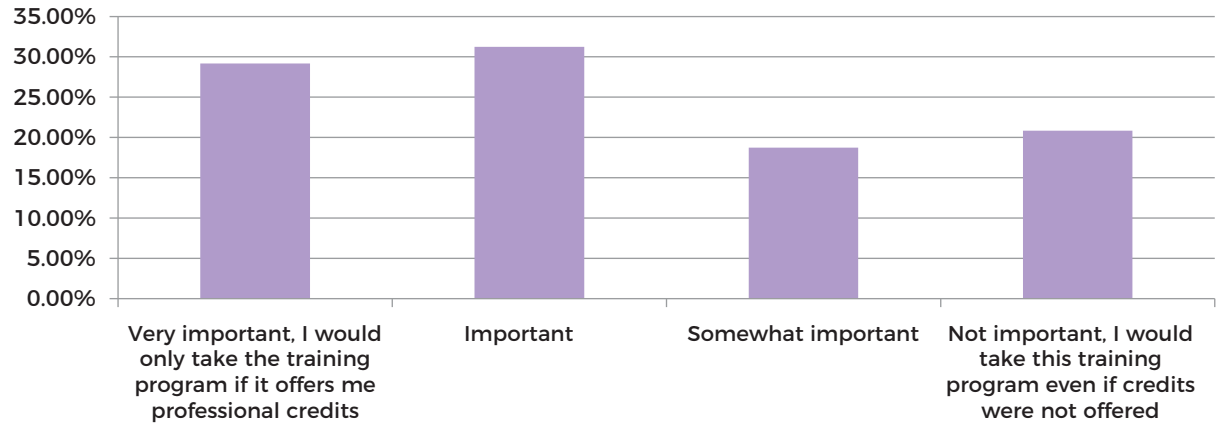
25. What would be your preferences for future learning activities?

n = 97



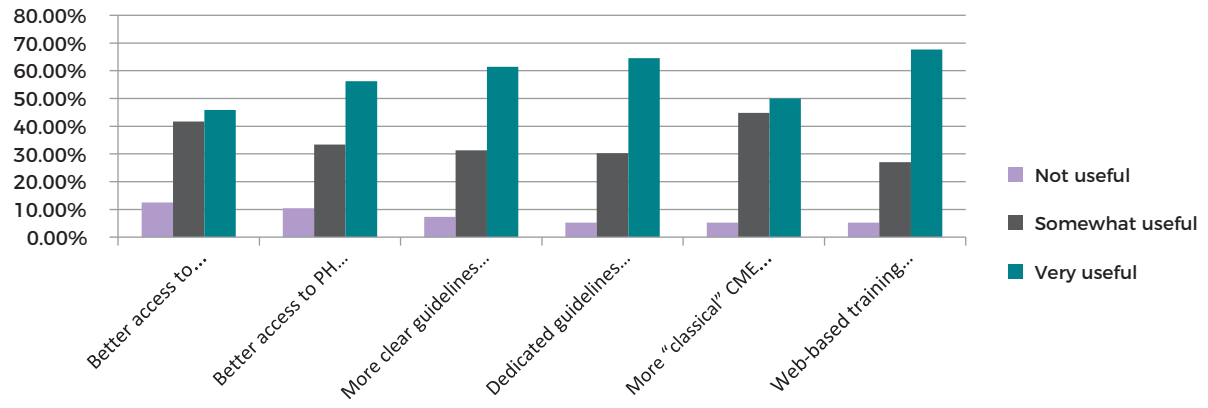
26. How important would it be for you to receive professional credits in order to participate in a training program i.e. accredited program?

n = 96



27. How useful would you find each of the following to better prepare you to help PH, or suspected PH, patients in your practice?

n = 96



28. Do you have any other suggestions to help you with the diagnosis and/or management of your patients suffering from PH?

- Need better resourced clinics with more nurses ANPs, clerical help and social work
- Thank you for offering all of the CME courses you have been over the past few years.
- awareness
- Guest speakers who work with PH patients and do it on a local level. Also webinars as it is difficult to get funding to travel to conferences
- Respiratory access for an ongoing problem
- When to refer to specialist, what type of workup would be needed, red flags, current therapies for management and how to monitor appropriately once back in the care of primary care providers
- Last time I was aware of PH was because of the Phen/fen link.
- primary care needs assistance in diagnosing or suspecting, with clear referral pathways needed
- Not thought of enough by prescribers/MRP to do anything with abnormal test results, to put the pieces together. Guidelines for primary care MRPs would be helpful
- Know what to do once a pt echo is confirmed PH. Often pt gets set up on home 02 from admission to hospital and maybe will see cardiologist but rarely the focus despite usually being severely hypoxic because of PH.
- Information about the role of oxygen therapy
- A pamphlet (Guide) on PH with examples and clear indications. With photos and color printing.
- A teaching guide that would be very useful for educating patients and other healthcare professionals.

APPENDIX 2 - PAH MANAGEMENT TOOLS AND RESOURCES

LIST OF RESOURCES:

Pulmonary Hypertension Association of Canada
PHA Canada Home Page - Pulmonary Hypertension Association of Canada

PHA Canada report
"The impact of Pulmonary Hypertension on Canadians"

Canadian Thoracic Society
The PHull PHollow-up: Optimizing PAH patient outcomes throughout the patient journey Improving Outcomes & CTEPH: Updated Guideline

Canadian Lung Association
Pulmonary Hypertension | Canadian Lung Association

European Respiratory Society
2022 European Respiratory Society Guidelines on the diagnosis and treatment of Pulmonary Hypertension

APPENDIX 3 – ONGOING CHALLENGES

Drawing from the comprehensive survey results, insightful interviews with PAH medical experts, and the invaluable perspectives of PAH patients, here is a list of ongoing challenges that were identified.

1. Diagnostic delays and under recognition

- Non-specific symptoms, so can it take years for proper diagnosis, delaying important therapies.
- Physicians do not investigate adequately SOB that is unexplained and does not respond to the usual therapies.
- Misdiagnosis happens often: confused with asthma, obesity, heart disease or other lung diseases for example.
- Timely availability of diagnostic tests varies depending on Province (e.g. Alberta easy for echo access, Qc difficult).
- Different interpretations of some results depending on the expertise of who is looking at them.

2. Inadequate Disease awareness, education, and research

- Lack of exposure to PH in medical school.
- Knowledge gaps about PH among primary care physicians and in rural area hospital and clinics.
- Not enough awareness/screening in certain high-risk populations (e.g. HIV, connective tissue disease, chronic liver disease, congenital heart disease).
- Lacking personalized medication. No targeted therapy exists for every patient, just give several drugs at once.
- Need medications that do not just delay disease progression but actual halt, reverse or cure.

3. Limited access to specialized care centers

- Geography: Specialized PH clinics for patients can be difficult to get to (must fly in or long drives) especially for patients coming from the north of Canada. Certain treatments are IV and patient must travel.
- Some provinces only allow access to a specialized PH center if referred by a specialist, not a GP.

4. Fragmented care coordination and follow up

- Primary care physician not good at communicating to PH centers about their patients.
- Quality of diagnostic tests can vary depending on where they are done (e.g. in small community settings).
- Not enough collaborations between different professional societies to raise awareness for diagnosis, treatment and management.

5. Barriers accessing treatment and therapies

- Unequal accessibility for treatment and management. Limited access for some depending on where you live in Canada and your insurance coverage.
- Depending on the province, the health ministry can make it difficult to prescribe and access certain PH medications.

REFERENCES

1. Canadian Cardiovascular Society and Canadian Thoracic Society. (2020, July). CCS-CTS Pulmonary Arterial Hypertension Statement. Retrieved from https://cts-sct.ca/wp-content/uploads/2020/07/FINAL_CCS_CTS-PAH-Statement-July-2020.pdf

2. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. European Respiratory Journal, advance online publication. https://doi.org/10.1183/13993003.00879-2022:citation[oaicite:0]{index=0}.

3. Mayo Clinic Staff. (n.d.). Pulmonary hypertension - Symptoms and causes. Mayo Clinic. Retrieved from https://www.mayoclinic.org/diseases-conditions/pulmonary-hypertension/symptoms-causes/syc-20350697

4. Pulmonary Hypertension Association of Canada. (n.d.). PH Centres. Retrieved from https://phacanada.ca/Living-with-PH/PHCentres

5. YouTube. (2023, May 15). Advances in Pulmonary Arterial Hypertension Treatments [Video file]. Retrieved from https://youtu.be/9VikmNywyAs

6. Mayo Clinic Staff. (n.d.). Pulmonary hypertension - Diagnosis and treatment. Mayo Clinic. Retrieved from https://www.mayoclinic.org/diseases-conditions/pulmonary-hypertension/diagnosis-treatment/drc-20350702

7. Pulmonary Hypertension Association of Canada. (n.d.). Treatment Options. Retrieved from https://phacanada.ca/What-is-PH/Treatment-Options

8. Evolving Landscape of Pulmonary Arterial Hypertension and Redesigning Pulmonary Arterial Hypertension Clinical Trials Task Force of the 18th Global CardioVascular Clinical Trialists Forum. The evolving landscape of pulmonary arterial hypertension clinical trials. Lancet. 2022 Nov 26;400(10366):1884-1898. doi: 10.1016/S0140-6736(22)01601-4. PMID: 36436527

9. Pulmonary Hypertension Association of Canada. (n.d.). Clinical Trials in Canada. Retrieved from https://www.phacanada.ca/research/clinical-trials

10. U.S. National Library of Medicine. (n.d.). ClinicalTrials.gov. Retrieved from https://clinicaltrials.gov/ct2/home

11. Healio. (2024, May 29). Adults with PAH lose around 8% of annual workdays due to inpatient, outpatient care. Retrieved from https://www.healio.com/news/pulmonology/20240529/adults-with-pah-lose-around-8-of-annual-workdays-due-to-inpatient-outpatient-care

The Canadian Initiative: Exploring Adult Pulmonary Hypertension report was developed in collaboration with a wide range of healthcare professionals with expertise in pulmonary arterial hypertension, as well as patient partners.

AN INITIATIVE OF

RESPIPLUS

IN COLLABORATION WITH

PULMONARY HYPERTENSION ASSOCIATION OF CANADA
L'ASSOCIATION D'HYPERTENSION PULMONAIRE DU CANADA

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