RESPIPLUS™ PRESENTS

NOVEMBER 2025

PULMONARY ARTERIAL HYPERTENSION IN CANADA: BRIDGING THE GAP BETWEEN INNOVATION AND ACCESS

RESPIPLUS

PULMONARY ARTERIAL HYPERTENSION IN CANADA: BRIDGING THE GAP BETWEEN INNOVATION AND ACCESS

NOVEMBER 2025

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.

This Canadian Pulmonary Hypertension Initiative and creation of this report was made possible by a sponsorship from Merck Canada inc. The sponsors did not have any role in the development, content and review of this report.

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

Copyright © 2025 RESPIPLUS™. All rights reserved.

TABLE OF CONTENTS

i Executive Summary

ii Team of Experts

01 Section 1 - Introduction

05 Section 2 - Disease Burden & Patient Experience

10 Section 3 - Health System Challenges

18 Section 4 - Therapies and Innovations

22 Section 5 - International Comparisons

24 Section 6 - Strategic Recommendations

28 Key Messages

29 Conclusions

30 References

32 Appendices

35 Additional Resources

Copyright © 2025 RESPIPLUS™. All rights reserved.

EXECUTIVE SUMMARY

Pulmonary arterial hypertension (PAH) remains a rare but life-threatening disease where scientific progress has outpaced system readiness in Canada. Once uniformly fatal, PAH is now treatable in the short term, yet Canadians continue to face long diagnostic delays, inequitable access to therapies, and major gaps in coordinated care.

Through expert interviews, patient perspectives, and national data, this report examines where Canada stands and what must change to ensure timely, equitable care for all patients.

Diagnosis remains the greatest challenge, patients often wait more than two years for confirmation, with frequent misdiagnoses and inconsistent access to essential tests such as echocardiography. Specialized centres provide timely, high-quality care, but many patients face delays reaching them due to limited awareness, referral inconsistency, and inequitable access across regions.

Recent advances, including the introduction of new therapies that target disease mechanisms rather than symptoms, mark a turning point in PAH management. Yet without provincial reimbursement, expanded infusion and lab capacity, and a strengthened workforce, innovation risks deepening inequities instead of closing them.

The expert panel identified six priorities for reform:

Ensure universal access to early screening tools and standardized referral pathways to shorten time to diagnosis.

Expand workforce capacity by strengthening nursing, pharmacy, social work, and sonographer support within PH centres.

Align federal drug approvals with provincial reimbursement processes to accelerate equitable access to new therapies.

Integrate the national PH registry into routine clinical practice and policy planning to support real-world data and quality improvement.

Strengthen education and awareness across all levels of care, supporting primary care, specialists, and patients alike.

Guarantee equitable access to diagnostics, therapies, and follow-up care for all Canadians, regardless of region.

Canada now has the therapies, expertise, and data to achieve world-class outcomes. This report calls on clinicians, policymakers, and patient advocates to work together to close existing gaps, transforming medical innovation into accessible, coordinated care for every Canadian living with PAH.

Copyright © 2025 RESPIPLUS™. All rights reserved.

OUR TEAM OF EXPERTS

Panel, collaborating organizations & RESPIPLUS team members

SCIENTIFIC PANEL

Jason Weatherald, MD, FRCPC I Respirologist I Panel Chair
Paul Hernandez, MCDM, FRCPC I Respirologist I CTS Representative
Nathan Brunner, MD, FRCPC I Cardiologist I CCS Representative
Alan Kaplan, MD, CCFP (EM), FCFP, CPC (HC) I Family Physician I Chair FPAGC
David Langleben, MD, FRCPC I Cardiologist
Jaclyn Lam, Sonographer Educator
Jamie Myrah, BA I Executive Director, PHA Canada
Beth Slaunwhite I Expert Patient Partner

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.

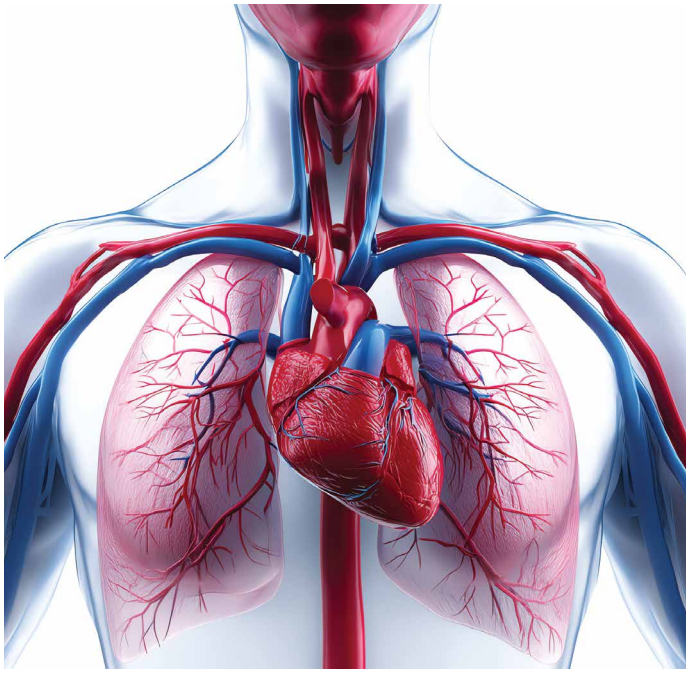
Maria Sedeno, MM, BEng | Executive Director
Emily Horvat, BSc | Senior Project Manager
Katrina Metz, RRT | Consultant, Medical Writer

CCS

The Canadian Cardiovascular Society (CCS) is the national voice for cardiovascular clinicians and scientists in Canada. Representing more than 2,500 members, the CCS works to advance heart health for all through leadership in research, education, and advocacy. The Society develops clinical practice guidelines, supports professional development, and fosters collaboration across the cardiovascular community to improve patient care nationwide.

Copyright © 2025 RESPIPLUS™. All rights reserved.

SECTION 1 - INTRODUCTION



Pulmonary arterial hypertension (PAH) remains a life-threatening and complex condition that exposes both the promise of medical innovation and the limitations of health system readiness in Canada. Over the past two decades, advances in therapy have transformed PAH from a uniformly fatal illness into a manageable disease (in the short term) for many patients.

Canadian outcomes appear broadly comparable to international reports but have shown limited improvement over time. Contemporary European registries describe 5-year survival of roughly 60–65% with current therapies *(Boucly et al., 2024)*. Population-based Canadian data *(Zelt et al. 2022)* report similar figures, approximately 89%, 76%, and 56% at 1, 3, and 5 years, respectively with a median survival near six years. While these datasets differ in design and patient selection, both point to a plateau in long-term survival despite advances in treatment. Persistent delays in diagnosis, inconsistent application of best-practice therapy, and regional variation in access may all contribute to this trend.

Recent therapeutic developments, including the approval of sotatercept in 2024 *(Product Monograph, CDA reimbursement recommendation, INESSS recommendations)*, signal continued progress in targeting the underlying biology of PAH. Yet this evolution also highlights a broader challenge: the need for a health system capable of adapting quickly and delivering complex therapies safely and equitably across regions.

This investigative report draws on expert interviews, patient perspectives, and supporting data to examine where Canada stands today, and what changes are needed to move from innovation to accessible, coordinated care for all patients.

Copyright © 2025 RESPIPLUS™. All rights reserved.

INTRODUCTION

1.1 BACKGROUND

Pulmonary arterial hypertension (PAH) is a rare but serious condition that exists within the broader spectrum of pulmonary hypertension (PH). In Ontario, analysis of more than 50,000 patients with PH showed that between 1993 and 2012, the annual prevalence rose from 99.8 to 127.3 cases per 100,000 people, while incidence increased from 24.1 to 28.7 per 100,000 between 2003 and 2012 *(Wijeratne et al., 2018)*. Most of these cases reflect other forms of PH related to left heart or lung disease, yet the overall rise underscores increasing detection and awareness of pulmonary vascular disorders. Within this broader group, PAH remains the rarest and most life-threatening subtype, requiring specialized investigation and treatment. Historically, the prognosis was dire without therapy, median survival was only less than three years after diagnosis, a fact that many patients still encounter when seeking information online. This stark reality explains the fear that often accompanies a new diagnosis. While modern therapies have transformed PAH into a more manageable chronic condition for many, extending survival and improving quality of life, these gains are not universal and depend heavily on early recognition and equitable access to advanced therapies *(Boucly et al., 2024; Zelt et al., 2022)*. Although patient numbers remain small, the burden on individuals, families, and health systems is substantial *(PHA Canada, 2021; PHA Canada & Canadian VIGOUR Centre, 2023)*.

The Canadian PH community has made substantial progress in recent years, with specialized PH centres, improved access to oral therapies, and growing adoption of combination regimens *(Hirani et al., 2020)*. Yet, inequities persist. Patients continue to experience prolonged diagnostic delays, inequitable access to therapies, and variable support depending on geography and resources *(PHA Canada, 2021; PHA Canada & Canadian VIGOUR Centre, 2023; Zelt et al., 2022)*. International peers, particularly France and Germany, have demonstrated that outcomes improve when national registries are embedded into routine care and linked to quality monitoring and reimbursement *(Humbert et al., 2010; Hoeper et al., 2013)*. Canada risks falling further behind without similar reforms, namely a coordinated national approach that embeds registry participation into routine care, links data to quality monitoring and policy planning, and standardizes access to diagnostics and advanced therapies across provinces.

Copyright © 2025 RESPIPLUS™. All rights reserved.

1.2 OBJECTIVES

The objective of this report is to provide a consensus-driven analysis of the state of PAH care in Canada. Specifically, the report aims to:

- Assess the lived experience of Canadian patients and caregivers.
- Identify system-level barriers in diagnosis, workforce, therapy delivery, and equity.
- Highlight international models that could inform Canadian practice.
- Provide actionable recommendations to ensure that scientific innovation translates into real-world benefit.

In this report, equity refers to whether patients across Canada can access timely diagnosis, specialist assessment, and funded therapies regardless of where they live. It captures variation in provincial funding decisions, inequitable access of PH centres, gaps in sonographer and echo resources, and the ability to meet monitoring requirements for modern treatments.

1.3 SCOPE OF THE REPORT

This report focuses on pulmonary arterial hypertension (PAH), distinct from broader categories of pulmonary hypertension (PH). While some findings may have relevance across PH groups, the analysis is targeted specifically at PAH, where therapeutic innovation is most advanced.

The scope includes the Canadian health system context, with comparisons to select international models. It incorporates perspectives from clinicians, patient leaders, and policy stakeholders, supplemented by registry data and published literature. The report does not attempt to provide detailed clinical guidelines; rather, it seeks to identify system reforms required to ensure readiness for innovation.

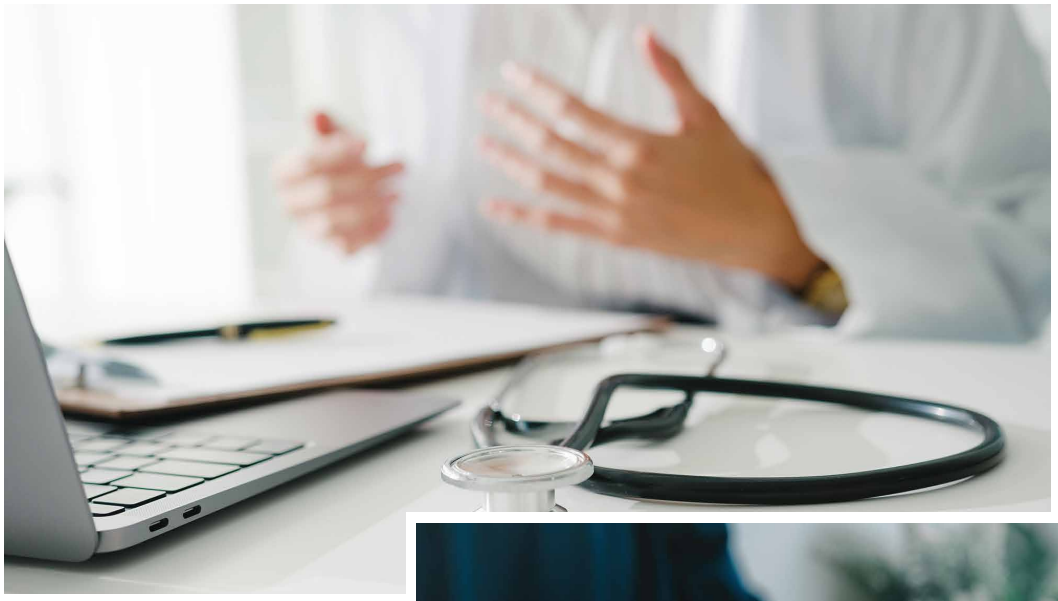
1.4 METHODOLOGY

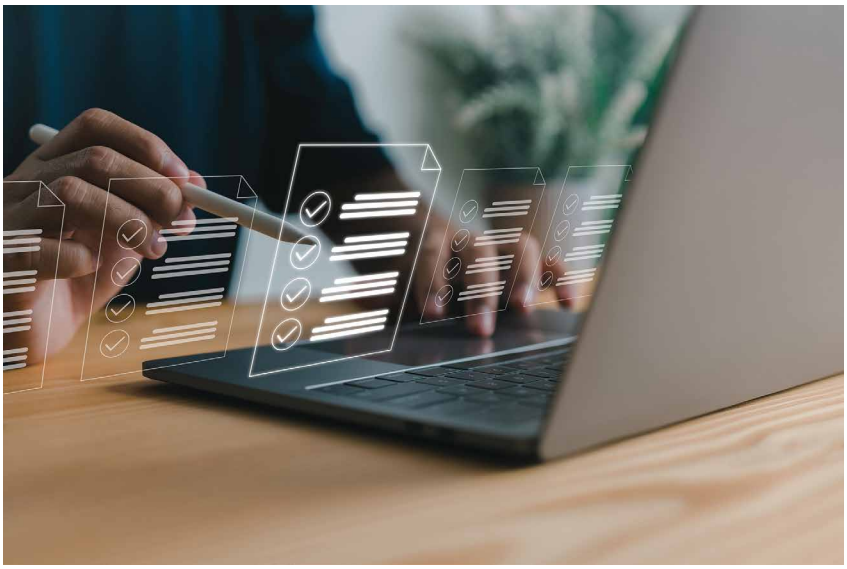
This investigative report was developed using a multi-source approach, with emphasis on expert panel interviews conducted in the summer of 2025. The panel brought together a broad cross-section of stakeholders, ensuring that clinical expertise, system perspectives, and patient voices were all included.

Interviews were semi-structured, enabling consistent exploration of diagnostic pathways, workforce needs, therapy access, registry integration, and equity. All sessions were transcribed and analyzed thematically. Direct quotations from participants are included throughout the report to illustrate key insights.

Copyright © 2025 RESPIPLUS™. All rights reserved.

Supporting evidence for this report was drawn from Canadian and international registry studies, trial data on emerging therapies, and published literature. To provide context specific to the Canadian experience, we also incorporated findings from prior surveys led by the **Pulmonary Hypertension Association of Canada (PHA Canada)**. The *2021 Canadian PH Community Survey (PHA Canada, 2021)* included more than 400 patients, caregivers, and health professionals across the country and highlighted diagnostic delays, treatment gaps, and quality-of-life impacts. The *2023 Socioeconomic Burden of PAH Survey (PHA Canada & Canadian VIGOUR Centre, 2023)*, completed by over 200 patients, provided detailed insights into employment, financial, and caregiver impacts. These surveys, designed and championed by PHA Canada, remain the most comprehensive patient-centered data sources available in Canada. Their findings were used selectively to enrich and contextualize the perspectives of the panel, but the primary emphasis of this report rests on expert consensus. See Appendix A for Methodology details.





Copyright © 2025 RESPIPLUS™. All rights reserved.

SECTION 2 - DISEASE BURDEN & PATIENT EXPERIENCE

Pulmonary arterial hypertension (PAH) is one of the rare diseases where the numbers seem small, but the consequences reverberate loudly across patients, families, and the healthcare system. In Ontario, population-based data show that the overall prevalence of pulmonary hypertension including all etiologic groups increased from 99.8 to 127.3 cases per 100,000 people between 1993 and 2012, with incidence rising from 24.1 to 28.7 per 100,000 between 2003 and 2012 *(Wijeratne et al., 2018)*. These findings reflect heightened detection and awareness of pulmonary vascular disease in general, rather than a true rise in pulmonary arterial hypertension, which remains rare. Survey data from PHA Canada (2021) and the Canadian VIGOUR Centre (2023) reveal that many people living with PAH experience profound losses in quality of life, employment, and independence, underscoring the social and economic burden of the disease The lived experiences captured in national surveys and patient interviews reveal a reality that is at once clinical, social, and deeply human.

2.1 EPIDEMIOLOGY AND NATURAL HISTORY

The epidemiology of PAH in Canada mirrors international trends. Roughly three-quarters of patients are female, most diagnosed between the ages of 30 and 60. Unlike many chronic conditions that affect older adults, PAH often strikes at a time of life when individuals are raising families, advancing careers, and contributing to the economy. This magnifies its social and economic toll.

Without treatment, median survival is less than three years. With current therapies, survival has improved substantially; Canadian registry data indicate 1, 3, and 5 year survival rates of 89%, 76%, and 56%, with a median survival of approximately six years *(Zelt et al., 2022)*. Contemporary European registries report broadly comparable outcomes, with 5-year survival typically around 60–65% *(Boucly et al., 2024)*. While differences across studies are modest and may reflect variations in patient selection, follow-up, or treatment era, the overall pattern suggests that long-term survival in PAH remains limited despite therapeutic advances. Continued improvements will depend on earlier detection, optimized treatment sequencing, and equitable access to specialized care

As Dr. David Langleben emphasized during his interview: “We have more drugs, but outcomes haven’t budged. That tells you it’s not only a science problem, it’s also systems problem."

Copyright © 2025 RESPIPLUS™. All rights reserved.

The natural history of PAH is one of inexorable decline. Patients describe a gradual but unrelenting narrowing of their world. At first, they can no longer keep up with friends on walks. Later, they struggle with stairs. Eventually, tasks as simple as showering or cooking become exhausting. This trajectory creates profound dependence and redefines the role of caregivers and family members.

International data confirm these observations. The US REVEAL registry demonstrated that without aggressive combination therapy, five-year survival remains below 60% *(Brown et al., 2011)*. French registry data demonstrate that outcomes improve when care is coordinated through national coverage and reference centres, supported by mandatory registry participation *(Humbert et al., 2010)*. In contrast, Canada's healthcare delivery remains highly regional, with voluntary and fragmented registry participation that limits its impact.

2.2 DIAGNOSTIC DELAYS

Diagnostic delay is perhaps the single most damaging aspect of the Canadian PAH experience. In the **2021 Canadian PH Community Survey**, half of patients reported waiting more than two years from the onset of symptoms to a confirmed diagnosis, while only 23% received a diagnosis within the first year. During those lost years, patients often endured repeated visits, inconclusive tests, symptomatic progression and mounting frustration.

"I was sent from specialist to specialist and told it was anxiety. By the time I finally got to a PH clinic, I could barely walk across the room."
Patient voice, 2021 Canadian PH Community Survey

"It wasn't just the shortness of breath, it was the constant not knowing, the feeling that nobody believed how sick I was."
Patient voice, 2021 Canadian PH Community Survey

"Unless there is a high index of suspicion in family practice, patients end up going down other pathways. We don't have systems that flag PAH early."
Dr. Alan Kaplan

Diagnostic delays remain substantial in Canada, often exceeding two years according to clinician reports and international comparisons *(Hoeper et al., 2013; Weatherald et al., 2018)*. Studies from the French PAH registry show a median delay of about one year when systematic referral pathways are in place *(Humbert et al., 2010)*. In the REVEAL registry, younger women were frequently misdiagnosed with anxiety or asthma before referral *(Brown et al., 2011)*, a pattern Canadian experts report still occurs, particularly in regions with limited access to echocardiography or PH specialists

Copyright © 2025 RESPIPLUS™. All rights reserved.

2.3 MISDIAGNOSIS PATTERNS

Misdiagnosis is not an accident, but a pattern rooted in systemic blind spots. According to the 2021 PHA survey, 41% of Canadian patients were first told they had asthma, 23% COPD, and 19% anxiety or depression. These typical misdiagnoses reflect the tendency of the system to default to common conditions rather than consider rare but serious alternatives.

The role of echocardiography is particularly important here. While echocardiography is widely available, sonographers often lack training in right heart assessment. These challenges are explored further in the section on diagnostic bottlenecks and workforce readiness.

This lack of training contributes directly to missed opportunities for early recognition. When echocardiograms are read without attention to right ventricular (RV) size and function, subtle signs of PAH are overlooked. By the time abnormalities are obvious, disease is often advanced.

The consequences of misdiagnosis are devastating. Patients spend months or years on inhalers or antidepressants that do nothing for their disease. They miss the opportunity for earlier referral and initiation of therapy. And once the correct diagnosis is finally made, the disease is often so advanced that outcomes are significantly worse.

2.4 SOCIOECONOMIC BURDEN

If the clinical burden is heavy, the socioeconomic burden is crushing. The **2023 Socioeconomic Burden of PAH Survey**, led by the Pulmonary Hypertension Association of Canada (PHA Canada) in partnership with the Canadian VIGOUR Centre, documented the profound impact on work and income *(PHA Canada & Canadian VIGOUR Centre, 2023)*. Nearly 90% of respondents reported occupational limitations due to PAH. Two-thirds said their ability to remain employed was compromised, and more than a third retired early. Another 29% reduced their hours or modified duties. Almost one in five were unable to work at all.

Copyright © 2025 RESPIPLUS™. All rights reserved.

The financial consequences of living with PAH extend beyond the disease itself. Although most advanced therapies are publicly covered, approval and renewal processes can be slow and inconsistent across provinces. While patients do not pay directly for these medications, many face other out-of-pocket expenses such as travel, parking, and lost income. Survey data from PHA Canada (2021) and the Canadian VIGOUR Centre (2023) highlight that these indirect costs and administrative barriers contribute to significant financial and emotional strain, even among those with formal drug coverage.

These burdens ripple into families. Caregivers frequently reduce their hours or leave employment to provide support, turning dual-income households into single-income, or no income ones just as expenses rise. In PHA Canada's Living with PH in Canada national survey, 43% of respondents reported receiving government financial assistance, and open-ended responses described financial insecurity despite long careers and previously stable household situations *(PHA Canada, 2021)*.

2.5 MENTAL HEALTH AND EMOTIONAL BURDEN

The psychological toll of PAH is overwhelming. In the 2023 survey, 88% of patients reported a negative impact on their mental health, and 38% described it as severe. Depression and anxiety were the most common conditions, but social withdrawal and fear of the future were also pervasive.

> Patient expert Beth Slaunwhite described the daily uncertainty: "Every day is filled with uncertainty. Will I be able to walk to the mailbox tomorrow? Will I still be here next year? That uncertainty wears you down."

> "As my wife's PAH has progressed, her need for a caregiver has increased exponentially. I have to go back to work but it's not easy finding an employer who will pay what I need to earn and allow the flexibility for me to attend her various appointments."
> Caregiver voice, 2023 Socioeconomic Burden Survey

International comparisons again highlight Canada's shortcomings. In France and Germany, psychosocial care is integrated into PH centre services. In Canada, it is often dependent on whether a centre happens to have a motivated nurse, social worker, or mental health professional willing to step outside their official duties.

Copyright © 2025 RESPIPLUS™. All rights reserved.

2.6 CAREGIVER BURDEN

Caregiving is central to the PAH experience but often invisible in policy discussions. The 2023 survey revealed that nearly half of patients required caregiver support. Among those with advanced disease, caregivers reported providing an average of 43 hours per week, essentially a full-time job layered onto their existing responsibilities.

Most caregivers were spouses or partners (62%), followed by adult children (15%). The remainder were parents, siblings, or friends. One spouse described the impact: “I work full-time, then come home to manage medications, oxygen, and appointments. It’s like having two jobs.”

The strain is not only physical but emotional and financial. Caregivers often sacrifice career advancement, retirement savings, and social connections. While most provinces have general caregiver support organizations, disease-specific programs for PAH remain limited, and few centres offer structured respite or counseling tailored to this population. Unlike in oncology or dementia care, caregiver support in PAH has not been systematized.

The impact of PAH on caregiver wellbeing is often overlooked, which in turn affects patient outcomes.. Exhausted and unsupported caregivers are less able to manage complex therapies or coordinate frequent appointments. Recognizing caregiving as a critical component of PAH care is essential for optimally improving clinical outcomes of patients with PAH.

2.7 SUMMARY

The burden of PAH in Canada is immense and multifaceted. Patients can endure long diagnostic delays, frequent misdiagnoses, and progression to advanced disease before receiving appropriate care. They face career disruption, financial strain, and profound psychological distress. Families provide extraordinary levels of unpaid caregiving, often at the expense of their own health and stability.

These burdens are not inevitable. International data show that earlier diagnosis, structured referral pathways, integrated psychosocial support, and recognition of caregiver roles can reduce suffering and improve outcomes. Canada has the expertise to deliver such care, but without structural reforms, patients and families will continue to shoulder a disproportionate share of the cost of this disease.

Copyright © 2025 RESPIPLUS™. All rights reserved.

SECTION 3 - HEALTH SYSTEM CHALLENGES

Canada's approach to pulmonary arterial hypertension (PAH) reflects both its strengths and its blind spots. On one hand, Canada has highly specialized pulmonary hypertension (PH) centres that deliver care on par with the best international institutions. On the other, patients often reach those centres late, and when they do, the workforce and infrastructure can be stretched thin. The result is a paradox: world-class expertise surrounded by systemic fragility.

3.1 DIAGNOSTIC BOTTLENECKS

The first and most persistent challenge is diagnosis. Canadian patients wait far longer than their peers in many other countries to receive a confirmed PAH diagnosis *(PHA Canada, 2021; Brown et al., 2011; Boucly et al., 2024)*.

As Section 2 described, half of all PH patients reported delays exceeding two years. This is not just an unfortunate statistic; it reflects structural weaknesses.

Limited biomarker coverage

Testing for blood levels BNP or NT-proBNP, key biomarkers of right heart strain, is not uniformly covered across provinces. In some regions, family physicians cannot order these tests without a specialist referral, creating a paradox where the very tool that could trigger referral is locked behind referral. This policy gap perpetuates late recognition.

Echocardiography inconsistency

While echocardiography is widely available, the protocols used by sonographers and cardiologists vary dramatically. Right ventricular (RV) dysfunction, a hallmark of PAH is often overlooked. Sonographer educator Jaclyn Lam stated: "Students are drilled on left ventricular function, but RV assessment is almost an afterthought. Many graduates simply don't feel comfortable flagging subtle RV changes." In addition, when abnormal right-sided cardiac pressures are detected suggestive of PH, they may be disregarded by both primary and secondary care physicians who are unsure how to interpret or act on the findings, when in fact such results should serve as an urgent red flag for further investigation.

Without standardized training and protocols, the same test may lead to different interpretations depending on the operator. This inconsistency undermines early suspicion and diagnosis.

Copyright © 2025 RESPIPLUS™. All rights reserved.

Access and Referral Pathways for Right Heart Catheterization

While echocardiography often raises suspicion of pulmonary arterial hypertension (PAH), a definitive diagnosis requires right heart catheterization (RHC). International guidelines recommend that RHC be performed at accredited pulmonary hypertension (PH) centres to ensure accurate and interpretable results. These centres are well established across Canada and, in most cases, can provide timely access once referral occurs. The challenge lies earlier in the pathway: community physicians may not always recognize when referral to a PH centre is warranted or may face unclear referral processes. Strengthening these referral pathways and ensuring PH centres have the resources to maintain rapid access are key to reducing diagnostic delay without compromising quality.

Dr. David Langleben emphasized: “Right heart catheterization is not just another test, it is the test. Without it, you cannot confirm PAH. Yet in many regions, patients wait far too long for what should be a basic standard of care.”

Dr. Nathan Brunner underscored the missed opportunity to engage cardiologists: “Cardiologists should understand when an echocardiogram may suggest PAH, and refer to an expert centre for further evaluation”

Fragmented referrals

Patients often cycle through multiple specialists before reaching a PH centre. Some see respirologists for presumed asthma or COPD, others see cardiologists for suspected heart failure, and many are told their symptoms are psychosomatic. Each referral adds months.

Dr. Brunner describes the referral problem here: “There is no standardized pathway between cardiology and PH centres. It depends on whether an individual cardiologist has experience or a personal connection to a PH program.”

3.2 WORKFORCE STRAIN

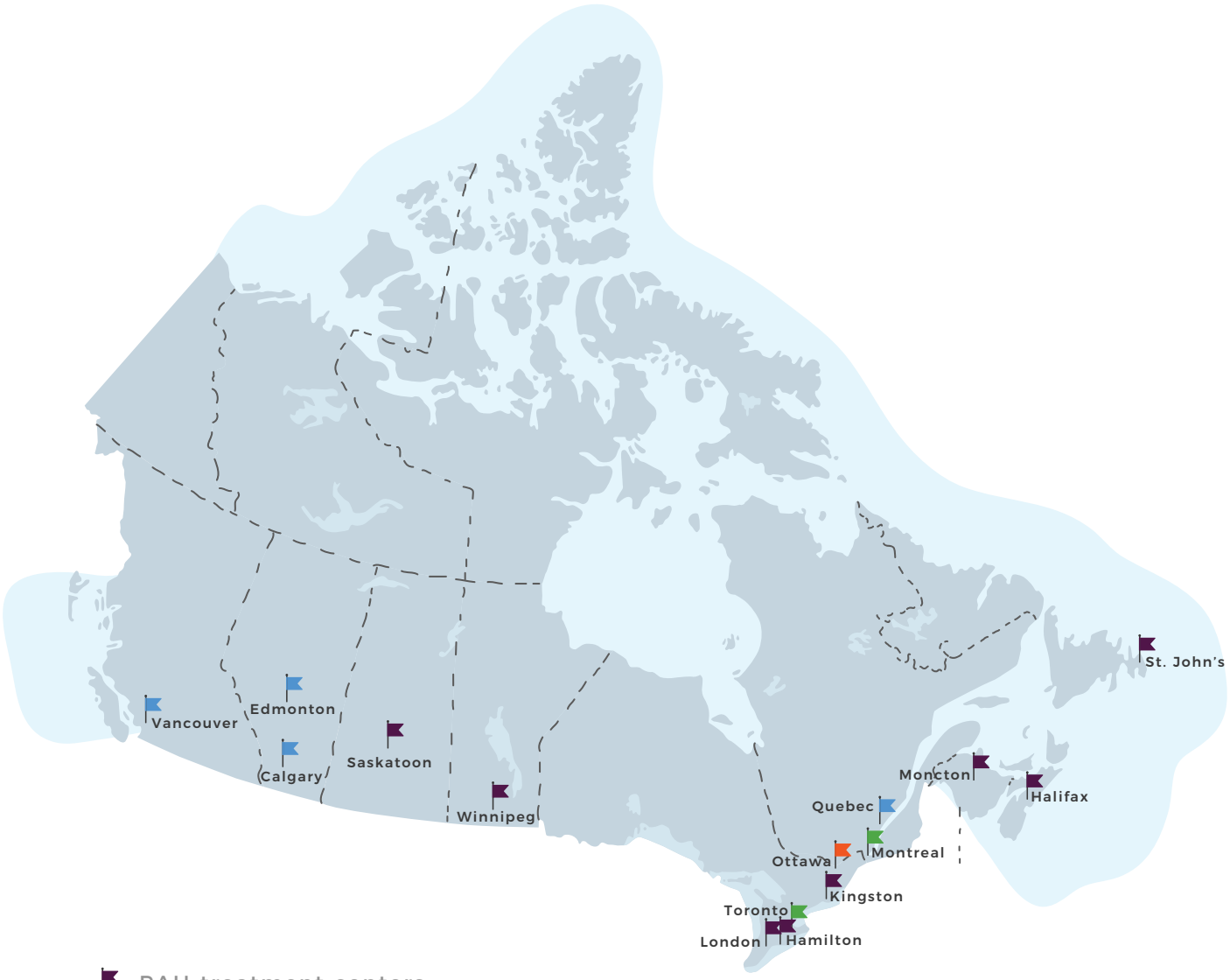
Once patients reach a PH centre, the quality of care improves dramatically. These centres are staffed by expert respirologists, cardiologists and nurses who have experience and specialize in PAH. These centres function as the hubs for advanced diagnostics, multidisciplinary care, and access to therapies. Across Canada, there are approximately 18 adult PH centres, located in major academic hospitals from Vancouver to St. John’s. A current list of centres is maintained by the Pulmonary Hypertension Association of Canada and can be accessed here: PH Centres.

Geographic access to specialized PAH care reflects Canada’s rural–urban divide. Most expert centres are located in major urban areas where patient density supports specialized services. However, patients in rural, northern, and remote regions face long travel distances for diagnosis and follow-up, particularly outside of central Canada.

Copyright © 2025 RESPIPLUS™. All rights reserved.

Centers across Canada

Below is a map of a specialized treatment centers across Canada



- 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

Copyright © 2025 RESPIPLUS™. All rights reserved.

However, the system's strength, specialist care of patients with PAH including prescribing medications, also underscores its weaknesses. Prescriptions for advanced therapies are appropriately limited to PH specialists, ensuring drugs are used safely and appropriately. Yet these specialists often work without sufficient allied health support, including nurse coordinators, social workers, and rehabilitation staff. Teams are in place, but their capacity is not aligned with the complexity of PAH care, which affects how fully they can deliver comprehensive, patient centred support.

Gaps in allied health

The key opinion leaders interviewed during the preparation of this report underscored the lack of nurses, pharmacists, social workers, and trained sonographers. Nurse coordinators are not adequately funded in PH expert clinics, and even if funded, they are stretched across large caseloads, leaving little time for patient education, ongoing therapy management, or monitoring. The demands extend beyond prostacyclin infusions, oral therapies also require dose adjustments, side-effect management, and regular lab reviews. Pharmacists, essential for optimizing complex regimens and ensuring safe drug interactions, are rarely embedded within PH centres. Social workers, who could help patients navigate the financial and emotional burdens of living with PAH, are often unavailable.

The sonographer challenges

Sonographers deserve special attention. As highlighted in Section 2, training programs do not emphasize PAH detection. This gap directly contributes to diagnostic delays. As Jaclyn Lam explains: "RV function was mentioned in lectures, but we never practiced it in labs. When I started in the clinic, I realized how unprepared I was."

Expanding sonographer education could accelerate detection across the country, but without dedicated funding or national standards, the gap persists.

Specialist overload

Specialists themselves are overwhelmed. With limited staff, they juggle diagnostics, therapy management, infusion oversight, and research obligations. Dr. Jason Weatherald explained: "We don't yet have infrastructure for biweekly monitoring or site-based follow-up. Without more nurses and allied staff, the system cannot absorb the next wave of therapies."

Copyright © 2025 RESPIPLUS™. All rights reserved.

3.3 THERAPY DELIVERY GAPS

Therapies for PAH are only as effective as the systems that deliver them. Canada faces persistent delivery gaps that undermine access.

Approval versus access

Health Canada approval does not guarantee availability. Provinces control reimbursement, which leads to significant delays between federal approval and public access. On average, Canadians wait about two years between Health Canada approval and provincial reimbursement for new drugs. Sotatercept's current status reflects this systemic reality: while approved federally in 2024, it remains unfunded provincially as of October 2025, consistent with Canada's usual timelines. The broader concern is not that this delay is unusual, but that such delays are routine for lifesaving therapies.

> Dr. David Langleben noted: "Approval means little if patients cannot access the therapy equally across provinces."

Infusion therapy capacity

Prostacyclin analogues remain lifesaving for high-risk patients, requiring continuous intravenous or subcutaneous infusion supported by trained nursing teams and patient education programs. While patient support programs for these therapies operate in all provinces, for patients in remote or rural communities, coordination of care and follow-up can still pose challenges despite national program support.

Monitoring infrastructure

New therapies, particularly sotatercept, require structured blood monitoring and close clinical oversight. Hemoglobin and platelet counts must be checked before the first dose and after the first five doses, with continued monitoring if results are unstable. After that, these labs need to be assessed periodically. Most centres are not prepared for this level of monitoring. Without laboratory integration and sufficient nurse capacity, therapies that could save lives risk being withheld for logistical reasons.

Copyright © 2025 RESPIPLUS™. All rights reserved.

3.4 REGISTRY UNDERUTILIZATION

Registries are the backbone of outcome improvement in rare diseases. France and Germany have demonstrated this repeatedly *(Boucly et al., 2024)*. Yet Canada's registry remains voluntary, incomplete, and largely research-oriented *(Zelt et al., 2022)*.

Lessons from France and Germany

In France, registry participation is mandatory and directly tied to reimbursement. No patient can receive advanced therapy without being entered into the registry. This ensures complete national data, allowing real-time outcome monitoring and policy evaluation. Germany similarly mandates reporting and uses registry data to inform cost-effectiveness reviews and therapy sequencing.

The US PHAR model

The Pulmonary Hypertension Association Registry (PHAR) offers the most current US model, with participation linked to PHA centre accreditation. The older REVEAL Registry is no longer enrolling.

Canada's missed opportunity

By contrast, Canada's voluntary registry provides incomplete data. Participation varies by centre, data entry is time consuming and not funded by clinical programs, and the absence of policy integration means the data do not shape reimbursement or care. This leaves Canada unable to answer basic questions: Are therapies improving survival nationally? Do outcomes differ by province or geographically within provinces?

Copyright © 2025 RESPIPLUS™. All rights reserved.

3.5 EQUITY ACROSS PROVINCES

Equity is perhaps the most glaring weakness in Canada's PAH care. Geography and provincial silos dictate access in ways that undermine fairness.

Provincial disparities

Access to PAH care and therapies remains inequitable across Canada. Some jurisdictions rely on lengthy special authorization processes for advanced therapies, while others have more centralized or structured review systems. In regions with limited specialist capacity or fewer designated PH centres, patients often face extended travel distances and longer wait times for diagnosis and treatment initiation. Even where programs exist, administrative requirements and workforce constraints can delay therapy start. Although every province provides a mechanism for high-cost drug access, the speed and ease of approval vary, making patient experience and time to treatment highly dependent on geography

> Patient expert Beth Slaunwhite summarized it powerfully: "Care is organized differently in every province, inequity is baked into the system."

Rural and Indigenous communities

Patients in rural and Indigenous communities face additional barriers. Travel costs, time away from work, and limited local diagnostic capacity all compound inequities. These patients may present with advanced disease and less likely to access the latest therapies quickly.

Financial inequities

Even when therapies are approved provincially, financial strain persists. **The 2023 Socioeconomic Burden of PAH Survey** revealed that 43% of patients relied on government assistance, and 10% skipped medications due to cost. These figures highlight how gaps in provincial funding and support programs continue to undermine adherence *(PHA Canada & Canadian VIGOUR Centre, 2023).*

Copyright © 2025 RESPIPLUS™. All rights reserved.

3.6 SUMMARY

Canada's health system challenges in PAH reflect a series of gaps that can undermine patient outcomes. Delayed diagnosis remains common, as family physicians lack consistent tools, biomarker coverage is inequitable, and sonographers are not systematically trained to detect right heart changes. When patients do reach PH centres, they receive excellent specialist care, but the centres often lack the nurses, pharmacists, and social workers needed to deliver fully integrated and optimal support. Federal approvals for new therapies do not automatically translate into provincial access, and the national registry remains underutilized for guiding policy. Inequities across provinces, rural and urban settings, and socioeconomic groups add further complexity.

Canada has the expertise and therapies to achieve world-class outcomes, but stronger systems are required to ensure that every patient can benefit. As Jamie Myrah of PHA Canada observed: "Families experience delays that can mean the difference between stabilizing disease and losing critical time. Equity shouldn't depend on your postal code."

Copyright © 2025 RESPIPLUS™. All rights reserved.

SECTION 4 - THERAPIES AND INNOVATION

Advances in pharmacology have transformed PAH from a uniformly fatal disease into one where patients can live longer and with improved quality of life. Patients across Canada have some inconsistent access to several classes of therapies, and combination treatment has become the global standard. Yet despite these innovations, Canadian patients continue to face inequitable access, delayed reimbursement, and infrastructure limitations that blunt the impact of new drugs.

4.1 CURRENT THERAPY CLASSES

Several classes of therapies are currently available in Canada, each addressing different pathways in the disease process.

Phosphodiesterase-5 inhibitors (PDE-5i):

Sildenafil and tadalafil were among the earliest PAH-specific oral therapies (Health Canada approvals in the mid-2000s). They enhance nitric oxide signaling to promote vasodilation. Delivered orally, they are generally well tolerated and are often used in first-line combination regimens. Coverage is available through provincial exceptional access programs, but administrative delays remain common.

Endothelin receptor antagonists (ERAs):

Bosentan (approved 2001), ambrisentan (2008), and macitentan (2014) block endothelin-mediated vasoconstriction and vascular cell proliferation. These oral agents are widely used, often in combination with PDE-5 inhibitors. They require liver function monitoring, which can add to complexity to care. Provincial coverage is usually available, but only through PH specialists.

sGC stimulator:

Riociguat was approved in 2014 as a soluble guanylate cyclase stimulator that enhances nitric oxide signaling. It is taken orally and is indicated for patients with inoperable or persistent CTEPH, and it also carries an approval for PAH. Access and reimbursement vary by province. In some regions, such as Alberta, it can be obtained through programs like STEDT, typically for PAH patients who have not responded adequately to PDE5 inhibitors.

Copyright © 2025 RESPIPLUS™. All rights reserved.

Prostacyclin analogues and receptor agonists:

Epoprostenol (approved 1997) remains the gold-standard therapy for advanced PAH. It requires continuous intravenous infusion via a tunneled central venous catheter. While life-prolonging, it imposes a high treatment burden. Patients report the difficulty of living with a catheter in their chest, constant pump management, and risk of recurrent, serious line-related skin infection and sepsis.

Treprostinil is available in subcutaneous and intravenous formulations. The subcutaneous form is effective but tolerance is often limited by infusion site pain. There are also oral and inhaled formulations of treprostinil but neither are approved in Canada.

Iloprost, an inhaled prostacyclin, provides a non-invasive option but requires frequent dosing (6–9 inhalations daily). Iloprost inhalation is not approved in Canada.

Selexipag is a selective prostacyclin receptor agonist, approved in 2016, provides an oral option targeting the prostacyclin pathway. It is effective in delaying disease progression and is used when parenteral prostacyclin is not feasible. Dose titration and tolerability can be limiting factors.

Despite these options, none of the established therapies significantly reverse the vascular remodeling that drives PAH. They primarily target vasoconstriction, endothelial dysfunction, and thrombosis.

Activin Signaling Inhibitors

The above established therapies target vasoconstriction, endothelial dysfunction, and thrombosis, and may also exert secondary effects on vascular remodeling, slowing disease progression and extending survival. As Dr. David Langleben explained: “These drugs keep patients alive longer and improve quality of life, but they don’t fundamentally change the biology of the disease.” While existing PAH therapies may influence remodeling indirectly through anti-proliferative and anti-inflammatory pathways, sotatercept, introduces a new mechanism of action by modulating the activin signaling pathway which is involved in vascular remodeling of PAH. This class represents a paradigm shift in the treatment of PAH, moving from symptomatic and vasodilator therapies to a potentially disease-modifying approach.

Sotatercept: Approved by Health Canada in 2024, sotatercept is the first-in-class activin signaling inhibitor, directly addressing abnormal vascular remodeling. Clinical trials (STELLAR, ZENITH, HYPERION) show significant improvements in functional capacity, hemodynamics, and reduced risk of clinical worsening events.

Copyright © 2025 RESPIPLUS™. All rights reserved.

4.2 COMBINATION THERAPY TRENDS

International evidence has made combination therapy the cornerstone of PAH management. The AMBITION *(Galiè et al., 2015)* trial demonstrated the benefit of upfront dual therapy, usually with a PDE-5i and an ERA, and high-risk patients increasingly receive triple therapy including prostacyclins.

Canadian practice is moving in this direction but unevenly. Access to dual therapy is relatively consistent, but escalation to triple therapy often depends on local expertise and infrastructure. Patients in smaller provinces may face longer delays or lack of infusions of capacity for system/clinic prostacyclins.

Dr. Jason Weatherald observed: “Combination therapy has transformed how we manage PAH, but in Canada, uptake is patchy. Patients in major centres are more likely to get triple therapy, while others wait or never receive it.”

Patient experiences reflect this inequity. In the 2023 socioeconomic burden survey, multiple respondents reported being kept on monotherapy despite deteriorating symptoms.

4.3 ACCESS TO NEW DRUGS

In 2024, Health Canada approved sotatercept, the first therapy to target vascular remodeling by modulating activin signaling. Clinical trials demonstrated significant improvements in exercise capacity, pulmonary vascular resistance, and reduced risk of clinical worsening. This approval was hailed as a major milestone. Dr. Langleben described its promise: “Sotatercept gives us a way to actually change the disease process, not just the symptoms.”

However, approval has not meant access. Provinces have not finalized reimbursement pathways, and infrastructure for monitoring, including labs every 3 weeks, and clinical oversight, is not yet in place. Dr. Weatherald emphasized this gap: “We don’t yet have infrastructure for site-based monitoring, which will be essential for sotatercept.”

As of October 2025, sotatercept remains inaccessible in Canada because provincial reimbursement has not been implemented, despite a positive CDA reimbursement recommendation in November 2024.

This perspective reinforces the central lesson of Canadian PAH care: scientific progress must be matched with system readiness.

Copyright © 2025 RESPIPLUS™. All rights reserved.

4.4 DELIVERY INFRASTRUCTURE NEEDS

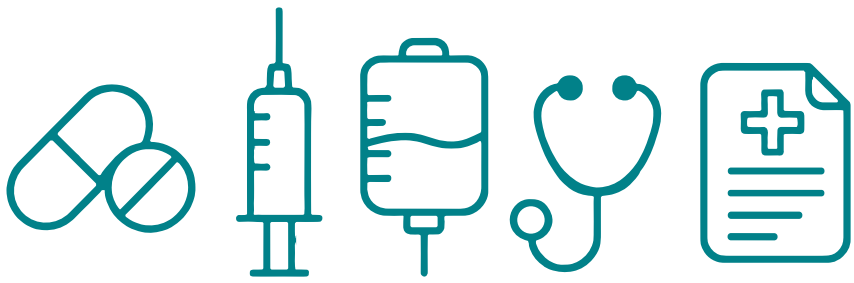
The success of PAH therapy depends as much on infrastructure as on pharmacology. Parenteral prostacyclins require infusion programs, sterile preparation, and dedicated nursing support. New therapies such as sotatercept require frequent routine laboratory monitoring and rapid coordination between patients, nurses, and physicians. Oral therapies are easier to deliver but still demand close adherence support.

Without workforce expansion and harmonized reimbursement pathways, new therapies will widen rather than reduce inequities. Canada risks creating a two-tiered system in which patients at large centres benefit from innovation while others wait.

4.5 SUMMARY

PAH therapy in Canada is at a crossroads. The available therapies offer unprecedented opportunities to improve outcomes, and new therapies may alter the natural history of the disease. But inequitable adoption of combination therapy, delays in provincial reimbursement, and limited infrastructure threaten to blunt these gains.

Therapeutic innovation alone is not enough. Without a healthcare delivery system that expands nursing capacity, integrates laboratory monitoring, and ensures equitable reimbursement, Canadian patients will continue to lag behind their international peers. The challenge is not just scientific but systemic, ensuring that every Canadian with PAH has timely access to the therapies that can extend and improve life.



Copyright © 2025 RESPIPLUS™. All rights reserved.

SECTION 5 - INTERNATIONAL COMPARISONS

International experience demonstrates that registry integration and system design make a measurable difference in outcomes. While Canada has world-class clinicians, its system is less structured than those of several peers.

5.1 FRANCE

France operates one of the most comprehensive national **PAH registries** in the world *(Boucly et al., 2024)*. Participation is mandatory, and data entry is directly tied to reimbursement for advanced therapies. This ensures near-universal coverage, enabling policymakers to track outcomes in real time and adjust access criteria accordingly.

Analyses of the French registry show improvements in survival compared to earlier eras, attributed not only to the structured use of registry data but also to more aggressive upfront use of parenteral therapies. By embedding accountability into access, France has achieved what Canada has not, a feedback loop between evidence, policy, and practice.

5.2 GERMANY

Germany's **COMPERA registry** is similarly robust, with mandatory reporting across specialized centres. Registry data are routinely used in cost-effectiveness assessments and to refine therapy sequencing. This integration ensures that advanced therapies are not only effective but also delivered efficiently *(Hoeper et al., 2022)*.

As Dr. David Langleben noted: "Other countries don't just collect data; they use it to shape practice and reimbursement. That's what Canada is missing."

Copyright © 2025 RESPIPLUS™. All rights reserved.

5.3 UNITED STATES

The Pulmonary Hypertension Association Registry **(PHAR)** is the key US registry, and participation is tied to PHA centre accreditation, which supports consistent data quality and benchmarking *(McLaughlin et al., 2025)*. The earlier REVEAL registry is no longer enrolling.

5.5 SUMMARY

International peers have shown that registries can drive outcomes when embedded into care and policy. Canada, by contrast, continues to treat its registry primarily as a research tool. Closing this gap is not just about data, it is about accountability, equity, and readiness for the next era of PAH care.

Copyright © 2025 RESPIPLUS™. All rights reserved.

SECTION 6 - STRATEGIC RECOMMENDATIONS

Canada has the clinical expertise and the therapies to transform outcomes in pulmonary arterial hypertension (PAH), yet system barriers persist. Drawing on survey data, registry evidence, and the perspectives of clinicians, patients, and advocates, the panel identified six areas for reform: diagnostic readiness, workforce expansion, therapy delivery, registry integration, equity and education.

6.1 DIAGNOSTIC READINESS

Timely diagnosis is critical to outcomes, yet many Canadians still wait years for confirmation of PAH. Delays reflect limited access to echocardiography, particularly outside major centres along with inconsistent use of supportive biomarkers such as BNP or NT-proBNP. As RHC should only be conducted by experienced teams, the goal is not to expand procedural availability, but to streamline referral and triage so that patients reach expert centres more quickly for accurate and safe hemodynamic assessment.

Variability in echo reporting standards and referral pathways further slows recognition. These gaps, compounded by workforce shortages and fragmented provincial funding, make timely diagnosis dependent on where patients live rather than on clinical need.

Expanding access to BNP/NT-proBNP testing, embedding right heart assessment into sonographer training, and implementing clear referral criteria from primary care would shorten diagnostic timelines. As Dr. Alan Kaplan noted: “Primary care physicians cannot suspect what they are not equipped to detect.”

Future projects could include continuing professional development programs for family physicians, targeted sonographer training modules, and awareness campaigns aimed at shortening the diagnostic journey.

Copyright © 2025 RESPIPLUS™. All rights reserved.

6.2 WORKFORCE EXPANSION

PH centres deliver excellent care, but they lack the allied health professionals needed to manage modern therapy regimens. Nurse coordinators, pharmacists, and social workers are either not funded or if so, then stretched thin, limiting the ability to provide comprehensive education, psychosocial support, and monitoring.

Investments in allied health staff would not only improve the delivery of current therapies but also prepare centres for the demands of new agents. Patient expert Beth Slaunwhite described the challenge well: “The care is excellent, but the teams are overworked. More staff would mean more time for patients.”

A practical step forward would be to explore hub-and-spoke models for regional support and pilot projects that fund allied health roles in PH centres.

6.3 THERAPY DELIVERY

Therapies cannot change outcomes if patients cannot access them. Health Canada approval is only the first step; national reimbursement and provincial funding processes often move too slowly, leaving patients without timely access. These delays reflect a system-wide bottleneck rather than provincial inequity. True inequities will emerge once provinces begin making different coverage decisions. The lag between sotatercept’s approval and its absence from public formularies underscores how long the system takes to translate scientific progress into patient care

Expanding medication infusion/injection capacity, strengthening laboratory networks for monitoring, and aligning reimbursement across provinces will be essential to ensure patients benefit from innovation. As Dr. Paul Hernandez cautioned: “New therapies must not widen disparities in access between provinces or patient groups.”

Future dissemination campaigns could focus on explaining reimbursement barriers, while RESPIPLUS and partner societies could lead educational webinars for clinicians to update them on new therapies, pathways, and patient access strategies.

Copyright © 2025 RESPIPLUS™. All rights reserved.

6.4 REGISTRY INTEGRATION

International peers have shown that registries are not just research tools but policy levers. France and Germany mandate registry participation, linking it to reimbursement and quality monitoring. Canada's voluntary registry remains incomplete, limiting its ability to shape decisions.

Embedding registry participation into routine care, and using the data to guide reimbursement and policy, would align Canada with international best practice. As Dr. David Langleben observed, without mandatory registry data, the system is "flying blind."

National societies could collaborate to expand education for clinicians on registry use and reinforce the importance of complete, high-quality data entry. PHA Canada already supports this effort through its dedicated webpage on the Canadian PH Registry and related educational initiatives for the PH community. Building on this foundation, the next step could involve joint advocacy with clinicians and policy groups to make registry participation a routine component of PAH care and a consistent element of quality improvement across centres.

6.5 EDUCATION

Sustainable progress depends on embedding PAH knowledge across the professional spectrum. From undergraduate medical education to continuing professional development, clinicians must be trained to recognize, diagnose, and manage PAH. From Dr. Brunner's perspective, structured education must extend to cardiologists: "The CCS could play a stronger role in raising awareness that PAH is not just a respirology disease. If every cardiologist learned to recognize red flags, referrals would be faster and survival would improve."

Other supporting medical education organizations such as RESPIPLUS can play a pivotal role by delivering webinars, accredited CPD modules, and case-based programs that translate guidelines into practice.

Copyright © 2025 RESPIPLUS™. All rights reserved.

6.6 EQUITY AND SYSTEM REFORM

Geography and provincial silos currently dictate access. Rural and indigenous patients face greater travel burdens, and provincial formularies create a patchwork of therapy availability. Harmonizing access criteria across provinces and expanding telehealth would reduce inequities.

As Dr. Paul Hernandez emphasized: “If PAH is to serve as a model for rare-disease policy, then we have to show Canada can deliver equitable access across provinces. Otherwise, we’re just adding to the patchwork.”

Targeted awareness campaigns could highlight specific disparities in care, such as the longer diagnostic wait times in some provinces, inconsistent access to echocardiography and BNP testing, and inequitable funding for advanced PAH therapies. Future projects could include advocacy-focused webinars and stakeholder roundtables aimed at reducing these provincial gaps and advancing coordinated policy reform.

Copyright © 2025 RESPIPLUS™. All rights reserved.

KEY MESSAGES

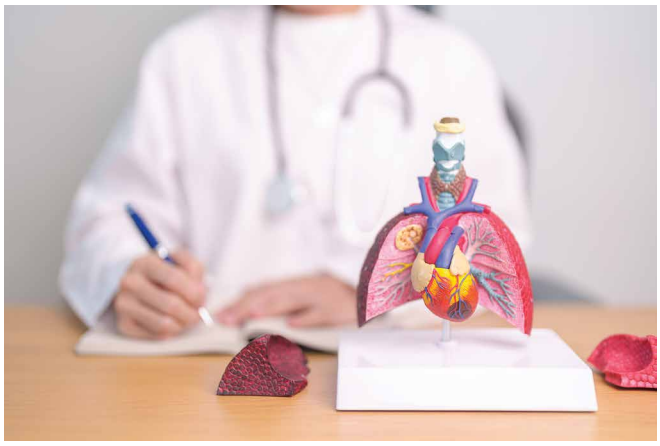
Early diagnosis saves lives
Patients wait too long for a PAH diagnosis. Universal access to biomarkers, standardized sonographer training, and clear referral pathways are needed to shorten delays.

Build capacity around PH centres
Specialist centres deliver excellent care, but nurses, pharmacists, and social workers are overstretched. Expanding allied health staff is essential for safe delivery of current and future therapies.

Approval must mean access
New therapies are approved by Health Canada but not yet available. Faster provincial reimbursement expanded infusion and lab capacity, and harmonized access criteria are required.

Data must drive decisions
Canada's registry is underused. Embedding mandatory participation and linking it to policy will enable accountability, equity, and continuous quality improvement.

Equity is non-negotiable
Rural and indigenous patients face greater barriers, and provincial silos create a patchwork of access. Harmonized pathways and expanded telehealth can reduce inequities.



Copyright © 2025 RESPIPLUS™. All rights reserved.

CONCLUSION

Pulmonary arterial hypertension shows both the progress of science and the gaps in Canada's system. We now have therapies that can extend and even reshape lives, yet patients continue to face long diagnostic delays, inequitable access, and inequities across provinces.

The priorities are clear: earlier diagnosis, stronger teams around PH centres, streamlined approval-to-access pathways, registry integration, and equity. These reforms are achievable and urgent.

With innovation at hand and expertise in place, Canada can lead the way in transforming PAH care and out patient outcomes.

Copyright © 2025 RESPIPLUS™. All rights reserved.

REFERENCES

- Balasubramanian A, et al. Health-related quality of life across the spectrum of pulmonary hypertension. Chest. 2024;165(6):1493–1504.
- Benza RL, Gomberg-Maitland M, Miller DP, Frost A, Frantz RP, Foreman AJ, et al. Predicting survival in pulmonary arterial hypertension: REVEAL risk score 2.0. Chest. 2019;156(2):323–37.
- Boucly A, Beurnier A, Turquier S, Jevnikar M, de Groote P, Chaouat A, et al. Risk stratification refinements with inclusion of haemodynamic variables at follow-up in patients with pulmonary arterial hypertension. Eur Respir J. 2024;64(3):2400197. doi:10.1183/13993003.00197-2024
- Brignole M, Moya A, de Lange FJ, Deharo JC, Elliott PM, Fanciulli A, et al. 2018 ESC Guidelines for the diagnosis and management of syncope. Eur Heart J. 2018;39(21):1883–948.
- Brown LM, Chen H, Halpern S, Taichman D, McGoon MD, Farber HW, et al. Delay in recognition of pulmonary arterial hypertension: factors identified from the REVEAL registry. Chest. 2011;140(1):19–26.
- Canadian Drug Agency. (2024, November). Reimbursement Recommendation: Sotatercept (Winrevair, Merck Canada Inc.). CADTH Submission SR0828. Retrieved from https://www.cdaamc.ca/sites/default/files/DRR/2024/SR0828REC-Winrevair.pdf
- Chin KM, Sitbon O, Benza RL, Badesch DB, Gaine S, Ghofrani HA, et al. Sotatercept in pulmonary arterial hypertension. Eur Respir J. 2024;63(6):2401325.
- Chin KM, Elwing J, Barst RJ, McLaughlin VV, Pulido T. Pulmonary arterial hypertension: epidemiology and registries. World J Hypertens. 2022;13(9):1699–1714.
- Dardi F, Elia D, Klinger JR, Coghlan JG, Rosenkranz S, Opitz C, et al. Advancing treatment paradigms in pulmonary arterial hypertension: lessons from registries and trials. Eur Respir J. 2024;63(4):2401234.
- Galiè N, Barberà JA, Frost AE, Ghofrani HA, Hoeper MM, McLaughlin VV, Peacock AJ, Simonneau G, Vachiery JL, Grünig E, Oudiz RJ, Vonk-Noordegraaf A, White RJ, Blair C, Gillies H, Miller KL, Harris JHN, Langley J, Rubin LJ. Initial use of ambrisentan plus tadalafil in pulmonary arterial hypertension. N Engl J Med. 2015;373(9):834-44. doi:10.1056/NEJMoa1413687.
- Hirani N, Swiston J, Lemarié C, Hirsch A, Mackenzie Ross R, Mehta S, de Perrot M, Lee L, Langleben D, Provencher S, Weatherald J, Hirsch GM, Granton J. (2020). Canadian Cardiovascular Society/Canadian Thoracic Society position statement on pulmonary hypertension. Canadian Journal of Cardiology, 36(7), 977–992. doi:10.1016/j.cjca.2020.04.030. PMID: 32654829.
- Hoeper MM, Huscher D, Ghofrani HA, Delcroix M, Distler O, Schweiger C, et al. The COMPERA registry: evaluating treatment strategies and outcomes in pulmonary hypertension. Eur Respir J. 2017;50(2):1602420.

Copyright © 2025 RESPIPLUS™. All rights reserved.

REFERENCES

- Hoeper MM, Huscher D, Ghofrani HA, Delcroix M, Distler O, Schweiger C, Grunig E, Staehler G, Rosenkranz S, Halank M, Held M, Grohé C, Lange TJ, Behr J, Klose H, Wilkens H, Filusch A, Germann M, Ewert R, Seyfarth HJ, Olsson KM, Opitz C, Gaine S, Vizza CD, Vonk-Noordegraaf A, Kaemmerer H, Gibbs JS, Pittrow D. (2013). Elderly patients diagnosed with idiopathic pulmonary arterial hypertension: results from the COMPERA registry. Int J Cardiol, 168(2), 871–880. doi:10.1016/j.ijcard.2012.10.026. PMID: 23164592
- Humbert M, Kovacs G, Hoeper MM, Badagliacca R, Berger RMF, Brida M, et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Respir J. 2023;61(1):2200879.
- Humbert M, et al. Epidemiology and pathophysiology of pulmonary arterial hypertension. Eur Respir J. 2023;61(2):2200879.
- Humbert M, Sitbon O, Chaouat A, Bertocchi M, Habib G, Gressin V, Yaici A, Weitzenblum E, Cordier JF, Chabot F, Dromer C, Pison C, Reynaud-Gaubert M, Haloun A, Laurent M, Hachulla E, Cottin V, Degano B, Jais X, Montani D, Souza R, Simonneau G. (2010). Pulmonary arterial hypertension in France: results from a national registry. Am J Respir Crit Care Med, 173(9), 1023–1030. doi:10.1164/rccm.200510-1668OC. PMID: 16456139.
- Lajoie C, Provencher S, Sitbon O, Badesch DB, Bresser P, D'Armini AM, et al. Combination therapy versus monotherapy for pulmonary arterial hypertension: a systematic review and meta-analysis. Lancet Respir Med. 2016;4(4):291–305.
- McLaughlin VV, Hoeper MM, Badesch DB, et al. Sotatercept for pulmonary arterial hypertension within the first year of diagnosis. New England Journal of Medicine.2025;DOI:10.1056/NEJMoa2508170.
- Merck Canada Inc. (2024). Winrevair (sotatercept) Product Monograph. Health Canada Drug Product Database. Retrieved from https://pdf.hres.ca/dpd_pm/00076926.PDF
- Pulmonary Hypertension Association of Canada. Canadian PH Community Survey 2021: National Report. Toronto: PHA Canada; 2021.
- Pulmonary Hypertension Association of Canada. Socioeconomic Burden of PAH Survey 2023: Final Report. Toronto: PHA Canada; 2023.
- Wijeratne DT, Lajkosz K, Brogly SB, Lougheed MD, Jiang L, Housin A, Barber D, Johnson A, Doliszny KM, Archer SL. (2018). Increasing incidence and prevalence of World Health Organization Groups 1 to 4 pulmonary hypertension: A population-based cohort study in Ontario, Canada. Circ Cardiovasc Qual Outcomes, 11(2), e003973. doi:10.1161/CIRCOUTCOMES.117.003973. PMID: 29444925; PMCID: PMC5819352.
- Zelt JG, McArthur E, Al-Omari M, et al. (2022). Mortality trends in pulmonary arterial hypertension in Canada: a population-based cohort study. Canadian Journal of Cardiology, 38(7), 1029–1037
- Zelt JG, Parsons JA, Manji RA, Weatherald J, et al. Survival in Canadian pulmonary arterial hypertension registry after 2015: no improvement despite combination therapy. Eur Respir J. 2021;58(5):2101552.

Copyright © 2025 RESPIPLUS™. All rights reserved.

APPENDIX A. METHODOLOGY

APPENDIX A

This report was built using a structured but pragmatic approach designed to reflect real conditions in Canadian pulmonary arterial hypertension care. The goal was to combine expert insight, credible existing data, and a focused review of guidelines and provincial access information. No new clinical research or surveys were conducted.

Expert panel discussions

A national panel of respirologists, cardiologists, family physicians, allied health professionals, and a patient leader met in June 2025. The meeting focused on diagnosis challenges, testing gaps, workforce pressures, and provincial variability. A second meeting will review the draft report before publication.

Individual interviews

Panel members participated in scheduled interviews in August 2025. These discussions explored diagnostic pathways, sonographer training, access to monitoring, treatment decision making, readiness for complex therapies, provincial disparities, and registry use. Interviews were recorded and verified for accuracy.

Targeted guideline and policy review

The team reviewed selected guidance documents including the WSPH 2024 framework, recent US consensus statements, CTS and CCS recommendations, and assessments from CADTH and INESSS. Provincial formularies and diagnostic access policies were also examined. This was a focused scan rather than a systematic review.

Existing surveys and community data

Findings were supplemented by PHA Canada's 2021 community survey, the 2023 socioeconomic burden survey, and previous RESPIPLUS reports from 2022 and 2023. These sources provided additional context on patient experience, diagnostic delays, and barriers to access.

Interview Guide

Interviews followed a common set of prompts. Topics included early recognition of PAH, access to BNP testing and echocardiography, variability in sonographer training, referral patterns to PH centres, provincial differences in workforce capacity, monitoring practices, and system readiness for new therapies with frequent laboratory and follow up needs. Panelists were encouraged to share clinical examples.

Copyright © 2025 RESPIPLUS™. All rights reserved.

APPENDIX A. METHODOLOGY

Analysis

Interview notes, panel summaries, and guideline findings were reviewed manually. Themes were grouped across participants and cross checked against provincial access information and selected international guidance. The analysis looked for areas where clinical expectations diverge from actual practice, especially in diagnosis, monitoring, and equitable access.

Limitations

The work draws on expert insight rather than new quantitative data. Some provinces without major PH centres are underrepresented. Provincial funding and diagnostic policies may change after publication. The analysis reflects a limited number of interviews and meetings, although participants are national leaders. Patient perspectives rely on existing PHA Canada surveys and the patient representative on the panel.

Integration into the Report

Findings from interviews, guidelines, and provincial access scans were combined to describe current practice, identify structural gaps, and inform recommendations. Statements of fact were cross checked against guideline documents, CADTH and INESSS assessments, and provincial formulary information.

Copyright © 2025 RESPIPLUS™. All rights reserved.

APPENDIX B. INTERNATIONAL REGISTRIES

This section highlights three major international PAH registries often used for benchmarking survival, risk stratification, treatment patterns, and system performance. These sources help provide context for how Canadian data compares.

France – French Pulmonary Hypertension Registry
https://www.orpha.net/en/disease/detail/182090

Germany – COMPERA Registry
https://www.compera.org/

United States – PHA Registry
https://phassociation.org/get-involved/participate-in-research/phar/

Copyright © 2025 RESPIPLUS™. All rights reserved.

ADDITIONAL RESOURCES

To support readers who wish to deepen their understanding of pulmonary arterial hypertension (PAH) and related care systems, we highlight the following curated resources:

Canadian and International Guidelines

- Canadian Thoracic Society (CTS) Guideline Library: https://cts-sct.ca/guideline-library/
- American Society of Echocardiography (ASE) Guidelines on Right Heart Assessment and Pulmonary Hypertension: ASE Guidelines

Educational Programs and Training

- RESPIPLUS Pulmonary Hypertension section within Chronic Lung Disease resources: Pulmonary Hypertension - All You Need to Know
- Expand Courses PAH Online Learning Program (RESPIPLUS & CTS collaboration)

Patient and Advocacy Resources

- Pulmonary Hypertension Association of Canada (PHA Canada): https://phacanada.ca/
- PHA Canada List of PAH/PH Specialty Centres: https://www.phacanada.ca/findaphcentre

Professional Societies and Networks

- Canadian Cardiovascular Society (CCS): https://ccs.ca/
- Canadian Society of Echocardiography (CSE): https://csecho.ca/

Copyright © 2025 RESPIPLUS™. All rights reserved.