



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

# CONNECTIONS

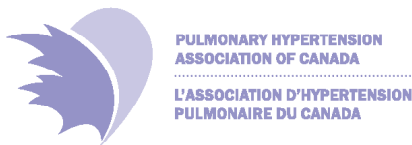
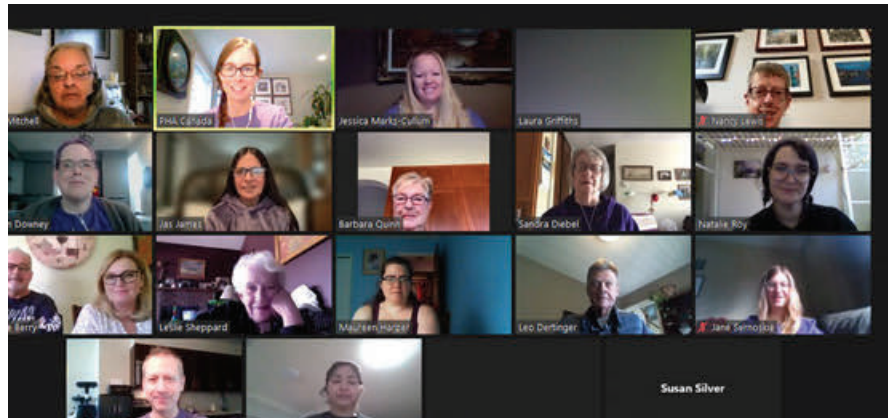
The Official Magazine of the Canadian PH Community



*Special Feature:*

## The Canadian PH Community Survey

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# Message from the Chair



## Resiliency Lives in All of Us

I can hardly believe I'm sitting down to write the message for this Winter issue. How did we ever get through another year of a global pandemic? When we celebrated PH Awareness Month in November, resiliency was the central theme, and it was appropriate in so many ways. I learned that resiliency lives in all of us, whether we think it does or not. The moment we wake up and start our day, we have no choice but to be resilient. I think it's important to remember that, especially for us PH patients.

We all know that living with a chronic illness, combined with a pandemic, is not exactly an easy thing to go through. So, there's the answer to my question: Resiliency! That's how we got through another year! Resiliency shines in all of us. In November, one of the quotes posted on PHA Canada's Facebook page was, "Sometimes carrying on, just carrying on, is the superhuman achievement" by Albert Camus. This quote resonated with me because it is the true definition of being resilient.

I think you'll really enjoy this issue of *Connections*. There are many great articles, including an interview with PH patient Derek Henderson, who served on our *Canadian PH Community Survey* Advisory Committee. I'm excited that we can share with you some of the preliminary results of the survey. I think you'll agree with me when I express how important it is to have data and information from our PH community. It is not only helpful to PHA Canada, but it helps physicians and researchers too. PHA Canada is so pleased with the response received by our survey—thank you for your participation.

Last year really exemplified just how amazing all your contributions have been. We raised a PHenomenal amount of money during PH Awareness Month, especially as part of the *6-Minute Walk for Breath*. We also had other fantastic fundraisers last year, including the annual *Go!PH for PH*. Fundraisers such as these allow us to award scholarships to research trainees working in PH. We know PH research is important to patients and their families. Recently, we were pleased

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[...] The moment we wake up and start our day, we have no choice but to be resilient

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to honour our late PHriend, Loretta Chu, with a scholarship in her name. None of this would be possible without the outstanding support and generosity of the PH community.

The PH community, albeit small, is a persevering, strong, generous, and resilient one. I do not doubt that 2022 will be another year where resiliency will shine through. I know the winter months can be more challenging for some patients who live in colder climates. I hope you will take the necessary precautions to be well, stay safe, and keep warm.

A handwritten signature in black ink that reads "Nicole Dempsey". The signature is written in a cursive, flowing style.

**Nicole Dempsey**  
Living with IPAH since 2013  
Board Chair, PHA Canada

# Message from the Executive Director



## Expanding Knowledge, Expanding Opportunities

Welcome to another issue of *Connections*! Like Nicole, I always find it hard to believe how fast time flies between issues. We are very excited to share our Special Feature on the *Canadian PH Community Survey* that PHA Canada conducted last year. Over 400 members of Canada's PH community—representing patients, caregivers, families, health care providers, and researchers—responded to the survey, giving us incredible insight into the current realities of a PH diagnosis in Canada and the community's needs in the future. Thank you to each person who took the time to complete the survey and share your thoughts, feelings, and experiences with us. Because of you, PHA Canada is better equipped than ever to lead the changes needed to improve the lives of Canadians affected by PH.

I also want to take this opportunity to thank the members of the volunteer Advisory Committee who helped PHA Canada build and design the survey (page 18). The Committee was tasked with prioritizing the many questions we wanted to ask and ensuring that the survey was user-friendly and would also gather the information most important to the PH community. It was an immense task that was beautifully accomplished thanks to the Committee's effort, expertise, and excellent editing. Bravo!

In this issue, you will learn more about what the results mean to patients (page 16), what we're learning about the journeys of PH caregivers (page 20), and who exactly completed the survey (page 24). We also provide an update on the Canadian PH Registry (CPHR) in response to survey results that show many patients and caregivers are still unaware of this important pan-Canadian initiative (page 22). You can also check out our webinar from PH Awareness Month, *Cultivating Resilience* (page 8), which shares information from the survey about the impact of the pandemic and the many ways you are practicing resiliency every day.

We will also introduce you to the latest recipients of PHA Canada's PH research scholarships (page 26). These \$10,000 scholarships are awarded to research trainees working in pulmonary hypertension, providing financial support for students and contributing resources to the academic centres conducting PH research. The projects that 2021's recipients—Yann Grobs (Laval University) and Pierce Colpman (Queens University)—are involved in demonstrate how exciting and innovative the PH research landscape is in Canada. Their projects investigate crucial mechanisms involved in the development of PAH and how to disrupt this development through novel treatment

strategies potentially. Understanding the underlying factors involved in PAH development is essential to diagnosing patients sooner and creating treatments that can stop or even reverse disease progression. The role of epigenetics in PAH development is further explored in this issue in a research update from 2018 scholarship recipient Dr. Francois Potus (page 28).

PHA Canada's support for PH research wouldn't be possible without the community's fundraising efforts. It has been incredibly heartening to witness the community's passion and commitment throughout the pandemic. Your support in 2021 for annual fundraisers like *GolPH for PH* (page 11) and the *6-Minute Walk for Breath* (page 10) exceeded our expectations and have inspired PHA Canada to set even bigger goals! Your gifts—both your donations and the time spent fundraising—are laying a foundation for PHA Canada to significantly increase our contributions to PH research in the coming years.

The *Canadian PH Community Survey* is just the tip of the iceberg. When combined with what we're learning through other national initiatives like the CPHR and the *Priority Setting Partnership* (a recent project led by Dr. Weatherald to establish community-led PH research priorities), our understanding of Canada's PH community is expanding as rapidly as our knowledge of PH. And as our knowledge grows, so do opportunities for patients to provide leadership in the design of everything from clinical trials to disease management programs, patient/family services, and more.

In 2022 we will be inviting patients to develop their skills and knowledge further so they can be meaningful participants in creating the changes needed. As always, PHA Canada will be with you every step of the way, providing the practical—and sometimes not-so-practical but still necessary—support you need to accomplish your own goals. Our purpose is to empower you to live better, so meeting you where you're at—wherever that may be from day to day—will always be part of our mission.

A handwritten signature in black ink that reads "Jamie Myrah". The signature is fluid and cursive, with the first letter of each name being capitalized and prominent.

**Jamie Myrah**  
Executive Director, PHA Canada

# MEMO: Inside PHA Canada

Welcome to *Connections'* message board. This is your peak into things happening inside PHA Canada, from changes in leadership to new resources, special events, and more.



## Retirement

### Emily Pinckard

Emily Pinckard joined the PHA Canada Board of Directors in May of 2019. In addition to her personal experience as a CTEPH patient, Emily also brought with her a legal background and volunteer experience in governance-related roles. Emily retired from the Board in December 2021. We thank her for her dedicated service; her passion, advocacy, and expertise will be missed.

## Welcome!

### Joan Paulin

We welcome Joan Paulin to PHA Canada's Board of Directors. Joan is a caregiver and longtime PHA Canada Ambassador who works tirelessly to advocate and educate for PH.

Joan's daughter Brooke was diagnosed with severe PAH in March 2014 at 24. She has always considered herself to be Brooke's primary caregiver. Yet, because Brooke remains very independent, her support has probably been more emotional than physical—one is no less important than the other! So, she thinks a better descriptor of her role in Brooke's journey would be as an advocate.

Joan's advocacy started very early—on day one with Brooke's medical team! She wanted to understand what medications were available to treat PAH (and was shocked to learn that drug access depends on where you live in Canada) and when new treatments might be available to PAH patients. In 2017, Joan became a PHA Canada Ambassador and served as an Ambassador until her election to the Board of Directors in December 2021.



### Elgin Lim

PHA Canada is pleased to welcome our newest team member, Elgin Lim, Manager of Operations and Development. Elgin brings a wealth of experience and expertise to his new role on the leadership team.

Having spent over 16 years in the HIV community health sector, Elgin happily joined the PHA Canada family in November 2021 as Manager of Operations and Development. With ample experience working in member-driven organizations, he has been a strong advocate of patient-centred care and program delivery.

His accomplishments include the creation of a peer navigation program recognized as a Leading

Practice by Accreditations Canada, a community dental clinic recognized with the Merit Award by the Canadian Association of Public Health Dentistry, and a successful collaboration with the University of British Columbia to provide cultural competency and HIV sensitivity education for senior social work students and future health-care providers.

Elgin is excited to start the next chapter of his career with PHA Canada. He hopes to bring his knowledge and experience in community health and non-profit organizational development to help those living with PH and their families.

**Ambassadors are champions of the PH community who seek to inspire others and facilitate collective action in support of a better life for those affected by PH. We are pleased to welcome two new Ambassadors who fit this definition perfectly: Joan Gibson and Jane Sernoskie.**

### Joan Gibson

Joan's daughter, Jane, was diagnosed with idiopathic pulmonary arterial hypertension in 2016 when she was 26 years old. "I am so thankful that she has the fabulous medical team at the PH Clinic at the Ottawa Heart Institute and strong support from her extended family and the Ottawa PH Support Group. I consider myself to be one of her biggest fans!", says Joan.



Having worked in the federal government for 30 years, Joan understands how the political system works. This helps her advocate for better and more accessible treatments for people living with PH. Joan also benefited from spending many years volunteering with the Parkinson Canada Association and other non-profit organizations. She is a firm believer in not reinventing the wheel and is always looking for ways to learn from other well-run organizations.

Joan is excited about being an Ambassador for PHA Canada and looks forward to helping the organization provide support for people living with PH.

### Jane Sernoskie

After receiving a rapid diagnosis of idiopathic pulmonary arterial hypertension, in November 2016, Jane made it her life's mission to pay it forward. She does this by raising awareness and funds, and advocating for affordable PH treatments for all. She is a kindergarten teacher by day and a PH advocate and awareness raiser by night.



Jane lives in Ottawa, Ontario with her bouncy black Lab dog Penny and handsome husband Craig. They love their life in the country and enjoy doing outdoor activities together and spending time with their family and friends.

Jane enjoys connecting with her PHellow PHighters and their PHamilies at the Ottawa Support Group, at PHA Canada's Monthly Meet-ups, on the Canadian PHriends Facebook page, during PH Buddy chats, and by reading their updates in The Pulse and Connections magazine. If you want to chat with Jane, feel free to send her a message.

## World PH Day, May 5, 2022

Held annually on May 5th, World Pulmonary Hypertension Day raises awareness of PH and celebrates the strength and resilience of the PH community worldwide. It is also an opportunity to highlight new developments in PH research and treatment.

Since 2014, thanks to the efforts of the PH community, monuments have been illuminated in periwinkle purple on May 5th from coast-to-coast. This has included such landmarks as: BC Place (Vancouver); Calgary Tower (Calgary); High Level Bridge (Edmonton); CN Tower (Toronto); Niagara Falls (Niagara); Ottawa Heritage Building (Ottawa); Olympic Tower (Montreal), and City Hall (Halifax)!

To learn more about World PH Day and how you can get involved go to [phacanada.ca/worldphday](https://phacanada.ca/worldphday)



# 2022 National PH Conference

May 28 - June 4  
Registration opens  
April 18th!

Learn more at  
[phacanada.ca/conference](https://phacanada.ca/conference)



Join the Canadian PH community once again for the National PH Community Conference. See friends and meet new people from across Canada. The 2022 PH Conference will again feature a full week of educational sessions, social activities, and special events.



The conference was wonderful. I am a patient of 11 years, and I came away with some great information and connections. Thank you!

– Participant, 2021 National PH Community Conference



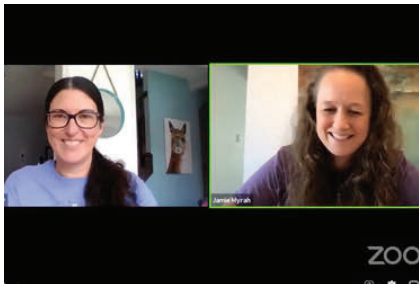
# Your Community

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Over the past year, the PH community has found creative ways to share experiences, raise awareness, and support each other, despite the challenges of COVID-19. You have shown resilience, strength, and compassion individually and as a community. This section is a celebration of your generosity. Enjoy highlights from November's PH Awareness Month activities, learn about the fantastic fundraising events organized and supported by the PH community, and read an update from Fondation HTAPQ in Québec.

# PH Awareness Month: Life in Purple

Throughout November PH Awareness Month 2021, the PH community from coast to coast to coast shone a bright light on pulmonary hypertension and what it is like to live “Life in Purple”. Despite continued COVID-19 restrictions, you shared your stories on how PH has affected your life through social media and our virtual events. Thank you so very much for your participation in our many virtual events including our Zoom Meet-ups, Facebook Lives, and *Cultivating Resilience* webinar. We are blown away by your support and dedication, which helped us raise over \$31,000 for the PH community. Thank you to everyone who participated in November’s events and helped make PH Awareness Month a success!

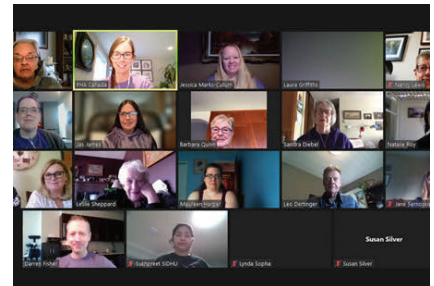


PHA Canada’s Board Chair Nicole Dempsey and Executive Director Jamie Myrah opened PH Awareness Month with a Facebook Live event talking about the importance of raising awareness of PH and inviting everyone to November’s events.

Milena Asghedom from PHA Canada and Line Ducharme from Fondation HTAPQ hosted a Facebook Live event talking about PHA Canada’s Monthly Meet-Ups and the value of connecting with others in the PH Community.



PHA Canada launched our PH Community Meet-Ups as a way for the PH community to stay connected during COVID-19. Since then, these virtual, peer-led gatherings have become an important place for patients and caregivers to meet others, all from the comfort of their own homes.



PHA Canada Vice-Chair Dr. Lisa Mielniczuk, from the Ottawa Heart Institute, and PHA Canada Ambassador Jane Sernoskie appeared in a feature interview with CTV Ottawa about PAH and the recently approved combination treatment Opsyvi® (macitentan and tadalafil).



Find webinar and event recordings, media coverage, and more community photos at [phacanada.ca/LifeinPurple](https://phacanada.ca/LifeinPurple)

Lynn Schnurr and her neighbour Diane in Kitchener, Ontario – Doing the *6-Minute Walk for Breath* with a friend!



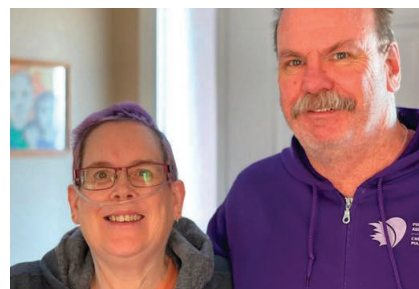
Tina Kirk in Calgary, Alberta shows off her “I am a PHighter” tattoo



Vanda McLean and her husband Gord celebrating in Orillia, Ontario



Kathy & Don Downey in Calgary, Alberta – Looking great in purple!



Jessica Cullum, Brin Marks & Brin’s Grandparents join the Walk from Sherwood Park, Alberta



Jane & Craig Sernoskie with Jane’s parents, Joan & Ian Gibson representing Ottawa, Ontario



The Chavarria family showing off their style from Vancouver, BC



Loretta Champdoizeau and Darold Deeter in Calgary Alberta, proud participants in the *6-Minute Walk for Breath!*



# 2021 Fundraiser Recognition

The PH community is filled with ambitious people ready to do whatever it takes to cure pulmonary hypertension. Hope drives them to overcome obstacles and find innovative ways to raise necessary funds for PH programs, including research. While the pandemic has certainly made community fundraising more challenging, it has also created new opportunities for people to support Canadians affected by PH.

We are incredibly grateful to the fundraisers who embraced the power of technology this past year and successfully raised thousands of dollars for PHA Canada. Your dedication to the cause allowed PHA Canada to award two more \$10,000 PH research scholarships in 2021 (see page 26) and helped us expand our support and education programs at a time when the PH community needs more support than ever. Here we honour 2021's most successful fundraising events: the *7th Annual GolPH for PH* and the *9th Annual 6-Minute Walk for Breath*. We can't wait to see what is in store for 2022, where we hope to see the return of other long-standing fundraisers like the *Ajax Walk/Run for PH Research* and *Delores's Heavenly Heart*.

## 6-Minute Walk for Breath

This year's *Annual 6-Minute Walk for Breath* in honour of PH Awareness Month was an amazing tribute to the resiliency of the PH community! Traditionally hosted in-person by the Ottawa PH Clinic, PHA Canada's signature fundraiser went virtual in 2020 and just keeps growing. Held on November 13, 2021, this year's event raised over \$31,000—an astounding new record!

We loved seeing photos from across the country as participants walked with their loved ones and showed off their *Life in Purple*. Thank you to everyone who made this year's *6-Minute Walk for Breath* so memorable. And a special congratulations to this year's top fundraisers!

To see photos of the *6-Minute Walk for Breath*, please go to page 9.

## Top Fundraising Teams

It was no surprise that this year's top fundraising team was the powerhouse group out of Ottawa, ON. Led by PH nurse Carolyn Doyle-Cox and PH patient Jane Sernoskie, the team raised over \$10,000! Thank you, Team Ottawa—you PHighters are Phenomenal!

In second place were the Mayville Movers, also from Ontario. This team came together to raise more than \$9,000 in honour of Nicole and Michael Mayville's five-year-old daughter, who was diagnosed with PAH in 2020. Way to go, Mayville Movers—what an amazing show of support!

Rounding out the top three teams was PHA Canada's own Board and Staff team. Together, this team of PH champions from across the country raised over \$4,000. Nice work, Team PHA Canada—keep up the good work!

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Help us find a cure for PH! Contact us  
at [donate @phacanada.ca](mailto:donate@phacanada.ca)  
to find out how we can help you raise critical  
funds for PH support, education, and  
research programs in Canada.  
—



## GolPH for PH

Although the Paulin family were unable to hold their annual golf tournament in honour of their daughter Brooke, who was diagnosed with PAH in 2014, their *GolPH for PH* supporters came out in full force and raised over \$17,000 for PHA Canada. It was a very special year of fundraising for the Paulins, as they were raising funds for a new PHA Canada Research Scholarship named for Loretta Chu.



Loretta Chu

Loretta Chu co-founded the Toronto PH Support Group in 2006 after being diagnosed with PAH at the age of 25. Loretta was committed to ensuring that newly diagnosed patients did not feel alone and became a tireless advocate for PH awareness and equitable access to treatments across Canada. In December 2020, as Loretta said her final goodbyes to the PH community, Darren Bell—PHA Canada Eternal PHriend and Founding Member—established the *Loretta Chu Memorial PH Research Scholarship* in her honour. Thanks to Darren and the fundraising efforts of the Paulin family, a \$10,000 scholarship was awarded in Loretta’s name in November 2021.



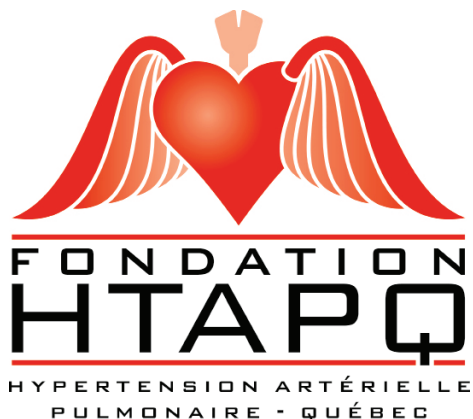
Jane & Family golfing

There may not have been the usual golf tournament, but there was still golfing! Jane Sernoskie and her family held a “mini-event” of their own in Ottawa, ON. Decked out in their PHA Canada gear, they enjoyed a round of golf and some lunch before making donations to *GolPH for PH*. That’s the spirit!



We will be eternally grateful for Loretta’s outreach in our darkest hours and are so happy to be given the opportunity to recognize her outstanding contribution to the PH community through a PHA Canada Research Scholarship in her name.

Pat & Joan Paulin



## Always Active!

2021 coincided with our Foundation's 15th anniversary. The Foundation continues its activities while respecting public health restrictions due to the pandemic. Thus, our financial assistance program, videoconference meetings, and fundraising campaigns kept our Board members and committees busy.

The book *Vivre avec l'hypertension artérielle pulmonaire (Living with Pulmonary Arterial Hypertension)* was published last year commemorating our Foundation's 15th anniversary. The book shares various stories of the Foundation—its birth, growth, and activities. The book also contains over 30 testimonials of people living with the disease, caregivers, and even transplant recipients who decided to share their journey. This publication promotes awareness of the disease, demonstrates its authors' resilience and, in addition, gives hope to those newly diagnosed. This book is for sale on the website of the HTAPQ Foundation. What a great way to raise awareness and promote early diagnosis!

The Foundation continues to receive requests for financial assistance from its members in need. A committee assesses financial assistance requests according to established criteria, such as the applicant's income and health.

Launched in 2020, we offer Essentrics classes to help those affected during the pandemic. Essentrics improves flexibility, strength, well-being and can even relieve chronic pain of participants. These are gentle exercises that are suitable for all fitness levels. Benefiting from the community's feedback, the trainer, Ms. Julie Bolduc, adapted the classes for people living with the disease. The Essentrics courses continues in 2022!

Our fundraising campaigns resumed after the easing of the pandemic health restrictions. Our Book Sale Committee organized a marquee

sale last September, where we sold thousands of new and used books. With the help of over 25 hardworking volunteers, we made an unprecedented profit. We raised over \$18,000, with expenses covered by generous sponsors.

In October, volunteers began preparing for the yearly Christmas fundraising campaign. We offered several choices to our loyal clients and friends of the Foundation: boxes of Québec cheeses, oil, and balsamic vinegar gift boxes, and our 15th anniversary book. Once again, the fundraising campaign was a huge success. It took more than 30 people to prepare the orders, and generous volunteers made deliveries across the province.

The Foundation pursues its mission: to improve the quality of life of people living with pulmonary arterial hypertension and their families through support and information. New projects will be finalized in 2022...

**Dolorès Carrier**  
Volunteer, HTAPQ Foundation

*Special Feature*

# The Canadian PH Community Survey

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Last year, we asked you to share your PH journey and personal experiences through the *Canadian PH Community Survey*. We are pleased and humbled by the overwhelming number of responses from those diagnosed with PH, caregivers, and healthcare providers. While each person answered the survey from their own unique perspective, collectively, the data provides an opportunity to understand the complexity of PH and its impact on the quality of life of different parts of Canada's PH community. In this section you'll learn more about the survey, including who filled it out and who was part of the survey Advisory Committee. You will also learn what the survey is teaching us about the caregiving journey and receive an update from the Canadian Pulmonary Hypertension Registry.

# The Canadian PH Community Survey: Sparking Change for a Brighter Future

Strengthening our understanding of your life with PH today so we can create a better life for all Canadians affected by PH tomorrow.

In 2013, PHA Canada conducted a Burden of Illness (BOI) survey of the Canadian PH community. The resulting analytical report of the survey data has been a crucial tool for educating decision-makers on the complex needs faced by families affected by PH and has been a powerful advocacy tool for many years.

To continue improving the quality of life of PH patients and caregivers, PHA Canada conducted a follow-up PH community survey in 2021 to measure the physical, social, financial, emotional, and psychological impacts faced by the Canadian PH community. Additionally, the survey captures relevant data that will allow us to evaluate PHA Canada's 2021-24 Strategic Plan and ensure we are succeeding in empowering the Canadian PH community through support, education, advocacy, awareness, and research. The survey will help us measure changes in how a PH diagnosis has impacted people since 2013 and provide new insights into critical areas. These include the quality of connections between people affected by PH, understanding of current treatment options, attitudes towards current/potential treatment options, priorities for educational and support resources, and community demographics.

As with all our initiatives, we recognized that we could not achieve our goals alone and relied on the meaningful participation of the PH community. Early in the process,

we formed an Advisory Committee consisting of PH patients, caregivers, and health care professionals. Throughout the Spring of 2021, they provided valuable recommendations and insights into the survey's development. Specifically, the committee provided advice on the survey's overall design, including content and format, reviewed drafts, and provided recommendations for changes and additions. Equally important, the committee provided the expertise necessary for ensuring that the questions asked were relevant and vital to the PH community and that the information gathered would allow for useful, evidence-based analysis.

The PH community came out in record numbers to have their voices heard. We had an overwhelming response and are sincerely grateful to everyone who gave their time to share their journey and experiences with us.

The result of this work was the creation of the *Canadian PH Community Survey*, which included three targeted surveys designed to capture the experience of people diagnosed with PH (patients), the family and friends who support them (caregivers), and the healthcare professionals that treat them. The survey launched on May 5, 2021 and was open for approximately two months. The survey was available in both English and French, promoted widely through multiple communication channels, and was accessible online or by phone. The PH community came out in record numbers to have their voices heard. We had an overwhelming response and are sincerely grateful to everyone who gave their time to share their journey and experiences with us.

As we analyze and share these results, we hope you will agree that the information gathered tells a richer and more complete story than ever before of how PH impacts people and how together we can achieve a better life for all Canadians affected by PH.

Contributed by: Darren Fisher, Strategic Initiatives Manager, PHA Canada



## Who responded



**305**  
Patients



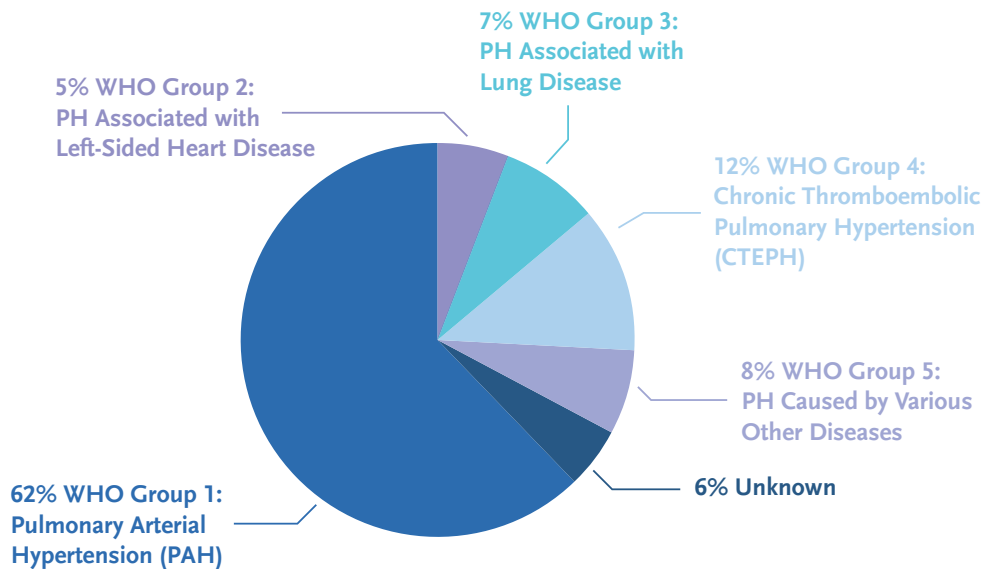
**77**  
Caregivers



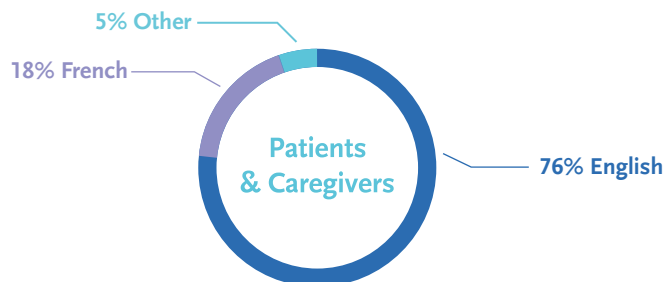
**24**  
Health Care  
Professionals

**Total = 406 responses**

## Type of PH reported by patients



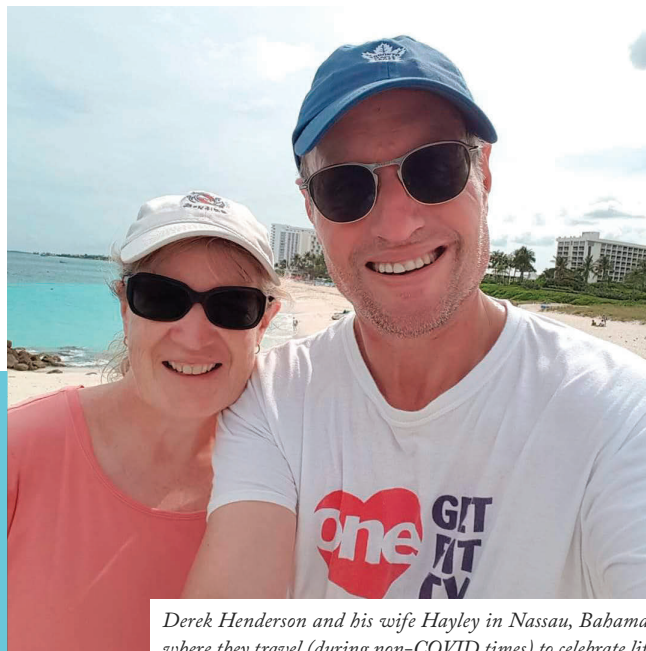
## Language



# Building the Survey: Bringing the CTEPH Perspective

PHA Canada recently spoke with CTEPH patient Derek Henderson about his experience as a member of the *Canadian PH Community Survey Advisory Committee*.

*This interview has been edited for length and clarity.*



Derek Henderson and his wife Hayley in Nassau, Bahamas where they travel (during non-COVID times) to celebrate life

## Q: Can you tell me a bit about yourself?

A: I live in Toronto. I'm 56 years old, and I was diagnosed with PH about seven years ago. I was diagnosed with pneumonia twice, and then after a two-year gap, eventually they found PH. My type of PH was CTEPH. I had PTE [pulmonary thromboendarterectomy] surgery about seven or eight years ago at Toronto General Hospital, and I'm on anticoagulants. I was fairly active before [my diagnosis] and since treatment I've been more or less back to normal life.

## Q: What was your role on the survey Advisory Committee?

A: I was the CTEPH member on the committee, so I brought that perspective to the committee. It's also a little different for me in that I know many of the committee members have medical backgrounds, and I do not. I'm a web developer—a coder. So, I have a different background in that sense and because I have a different kind of PH. So, my role was to make sure the committee heard from the CTEPH perspective and to help test the final design and help ensure it would work across all platforms.

## Q: Why did you get involved with the survey committee?

A: When I was diagnosed with CTEPH, I reached out to a woman in the U.S. named Amanda. She was someone with CTEPH and because it was 7-8 years ago, it was a little difficult to get information about the disease. So, when I found out I was going to have the PTE surgery, I reached out because there weren't many people you could call or ask about what the experience was like, what it would do, etc. One thing she asked me to do in return was to help anyone I could on their PH journey.

So, I certainly want to help people by conveying the CTEPH experience. When you're diagnosed with PH, it's as if you need to learn a new language—the treatments, what blood counts are, the six-minute walk test, the heart echocardiograms, etc.—and it can be scary.

Doctors and nurses do the best they can, but they're under tremendous pressure to see as many people as possible. And sometimes it's nice to talk to somebody, not on the treatment team, but that's been through this experience. So, to me,

the survey is a way to help solicit the information that patients need so that PHA Canada can act as a sounding board, advocate, or a source of information for people that are not directly related to their medical treatment group but more as a peer group to help patients understand what the PH community means, and that's what the survey brings.

## Q: So, for you, the aspect of support on the human level is the most important aspect of the survey?

A: Yes. And also, to help people within different types of PH and to find out what resources people need when diagnosed so that the next group of patients have that important information—even if it's just life hacks you can use after surgery or the right questions to ask the doctors.

I know when I was diagnosed, I was overwhelmed, wondering, what does this all mean? I didn't know what "blood clots" meant. Because it's a rare disease, most people don't know about PH, let alone CTEPH, so it helps to have a peer group to help you understand what these things are.

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The survey is a way to help solicit the information that patients need so that PHA Canada can act as a sounding board, advocate, or a source of information for people that are not directly related to their medical treatment group.

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## 10% of patients who answered the 2021 Canadian PH Community Survey were diagnosed with CTEPH

29% received single oral PH therapy

55% received pulmonary thromboendarterectomy (PEA) surgery

16% received balloon pulmonary angioplasty (BPA)

75% had their PH persist after having PEA surgery

### **Q: What was participating on the survey committee like?**

**A:** The approach of the committee was great, including the variety of people on the committee—the physicians, patients, and caregivers—it was a free and open space to talk, and everyone was trying to figure out the best approach to the survey. A lot of work went into it. So, it was a good experience for me, even though I don't think that I contributed that much. But it's been useful to see the types of information people are gathering and what will be useful down the road as well.

### **Q: Why do you think the survey is important to the PH community?**

**A:** It's important to the community because it will help develop programs and improve the PH literature, and maybe even some courses or education, so it will help clarify what is needed for education and support. And the survey also helps advocacy groups so you can go to the physicians and governments and highlight the problems people with PH are having. The survey also allowed patients to provide PHA Canada with some guidance on what they need to develop, what's missing in their literature, what's coming up, and even how the coronavirus pandemic is changing everything—including how everyone is coping with it.

### **Q: How do you feel about the questions about the pandemic included in the survey?**

**A:** It's a snapshot in time, and I don't know if it's a bad thing or a good thing. The pandemic has advanced telemedicine, and I think that's sorely needed. I've seen a lot of people travelling to the PH clinics in Toronto, but hospital restrictions during the pandemic forced the governments to implement telemedicine, which they were really reluctant to do before. So, in that sense, there have been some good aspects. But of course, COVID-19 has limited some diagnostics and limited the human contact patients can have when seeking treatment. For example, if someone is having PTE surgery, they may not be able to have hospital visitors after surgery. There may also be no ICU [intensive care unit] beds available, in which case the PTE surgery would have to be cancelled. The survey captured a snapshot in time, of history, that we're all living through.

### **Q: PHA Canada aimed to create a diverse committee with people from different perspectives. Did you feel that this goal was achieved and why does having a diverse committee matter for the survey?**

**A:** With a diverse committee, you get all sides of the table. My wife had a critical illness a few years ago, and I've had a critical illness, so I've had both the experience of being a patient and a caregiver. The more well-rounded a committee is, the more opinions you have, which is a good thing. PHA Canada did a very good job getting as many diverse voices as possible—I mean, with PH, you're dealing with a very small subset of people to begin with and there aren't many volunteers available—so, even though the number of volunteers you can get involved in the survey is small, I think PHA Canada did a good job of getting as wide a variety of perspectives as possible.

### **Q: What did you enjoy most or what impacted you the most during this process?**

**A:** Just hearing all the different opinions and seeing that there's a lot of emotion connected to these rare diseases, where some people may not understand what you're going through. And that experience was conveyed through some very strong advocacy. Because you're usually dealing with PH alone, I didn't know about some of it and learned new things about the experience of other PHers. There's definitely a lot of passion for this work in the PH community and that was one of the things that fascinated me. I enjoyed connecting with people who have PH, even if CTEPH is different than other forms of PH. It's still the same sort of experience that everyone is living through, just in different forms.

Contributed by: Miriam Bergeret, PHA Canada Knowledge Translation Volunteer, interviewing Derek Henderson

# Canadian PH Community Survey Advisory Committee

To advise us in the planning and design of this important initiative, PHA Canada was fortunate to have a group of people willing to volunteer on an advisory committee. Membership for this committee included people from diverse backgrounds, expertise, and experiences. We thank each of them for their insight and commitment. To help introduce you to the people behind the survey's design, we asked the committee to share their thoughts with us on the question: *“Why does the Canadian PH Community Survey matter to the PH community?”*

**Beth Slaunwhite, living with PAH since 2016, Halifax, NS**

“This wonderful, collaborative effort will help clinicians and researchers learn patient preferences, shape research moving forward, and aid key decision makers in determining the type of resources needed for patients and caregivers affected by PH. Our input on disease management is so important, and with the amazing number of responses from across this large country, this survey will ensure those voices are heard.”



**Derek Henderson, living with CTEPH since 2012, Toronto, ON**

“The survey is a tool that helps identify what strategies are working for physicians and patients dealing with PH and identifies some of the gaps where resources need to be applied. It also provides an historical snapshot of how the PH community is doing during the pandemic.”



**Dr. Lisa Mielniczuk, Director of the Ottawa PH Clinic and Vice-Chair, PHA Canada, Ottawa, ON**

“This survey represents the first of its kind in Canada and provides insight and depth into our community in ways we have not previously explored. From this survey we will gain a better understanding of the challenges, priorities, and opportunities for our patients and caregivers living with PH.”



**Joan Paulin, PH Caregiver since 2014, Director, PHA Canada, Mississauga, ON**

“The questions that most interested me were the ones that asked about the awareness of and the willingness to participate in Canada's PH registry, clinical trials, and blood sample collection because the answers to these questions, as well as the quality-of-life questions that go beyond the types of questions typically asked in a clinical setting, will help to lay the groundwork for a more patient-centered approach to healthcare.”



**Lisa Kolkman, Nurse Practitioner at the Vancouver PH Clinic and member of PHA Canada’s Canadian PH Professionals Network, Vancouver, BC**

“The research we participate in usually focuses on pulmonary hypertension from our perspective. It is so important to hear from patients directly about what matters to them.”



**Michael Robach, Director, PHA Canada, Vancouver, BC**

“The results of this survey will provide us with a good snapshot of the progress we’ve made here in Canada in the last eight years. But they’re also fundamental in informing how PHA Canada can continue to support our community, and the areas of work that will require our attention and focus going forward.”

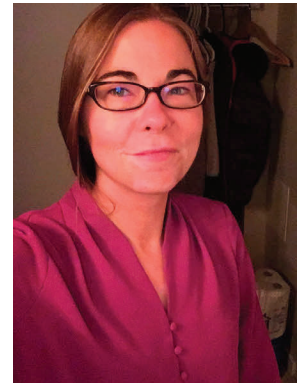


**Miriam Bergeret, living with PAH since 2016, Toronto, ON**

“As Peter Drucker said, ‘If you can’t measure it, you can’t improve it.’ That’s exactly why PHA Canada’s PH Community Survey is so important. The survey arms us with the knowledge and data we need to help improve the quality of life of Canadians with PH, to develop better resources for patients and clinicians, to raise awareness of PH in Canada, etc.”

**Tarya Morel, living with PAH since 2012, Vancouver, BC**

“It’s so important for patients and caregivers to have a voice in PH advocacy and treatment. This survey was about harnessing the collective voices and experiences of our community, so that our needs, our struggles, and our strengths can be shared back with clinicians, researchers, advocates, and back to the community as well. This information can be used to make life better for all of us, and I’m so pleased to have been a part of it.”



**Dr. Jason Weatherald, Director of the University of Alberta PH Program and member of PHA Canada’s Canadian PH Medical Committee, Edmonton, AB**

“I think the survey is important because it tells us how people with PH are doing across the country and provides insights into how things have changed over time. The information is rich and reflects many things we don’t get to touch on in the clinical setting. This survey also explores the unique experiences and challenges of living with PH in the current climate of COVID-19, which will be very useful going forward.”

# The Ongoing Story of You – The Resilient Caregiver

**Teri Kingston** joined the PH community in September 2012 when her husband, Harry Kingston, was first diagnosed with PAH (pulmonary arterial hypertension). She is a co-founder of the Ottawa PH Support Group and a former PHA Canada Ambassador. She has contributed several articles to *Connections* over the years. Her latest, in Fall 2019, details some of the lessons she learned after Harry passed away and her journey as a PH caregiver came to an end. Here she reflects on what the *Canadian PH Community Survey* has to say about PH caregivers and stories they have to tell.



Teri Kingston

Earlier this year, PHA Canada requested the PH community complete the *Canadian PH Community Survey*. As caregivers, you represented an important part of this community and generously shared your story, along with patients and healthcare professionals. Analyzing the results from patients, caregivers, and healthcare professionals collectively will provide valuable insights. Looking at the results provided by caregivers on its own is also a valuable exercise in its own right.

There are two ways to look at answers and data that come out of survey questions that are deep, broad, and wide. One is to analyze the data from a factual, objective standpoint; PHA Canada asked you specific questions, you gave specific answers. When these answers are summarized, charted, and graphed, we get a picture of what has happened to our community. The analytical process makes sense of the data. It provides decision-makers, executive members, and stakeholders with the evidence base they need to remain strategic and purposeful in managing this nationwide association for the benefit of all of us.

The second way to use the time, effort, and energy you have invested in this survey is to mine the stories peeking out of those graphs and charts. Or as data expert, Nancy Duarte says, “Oh, the stories data would tell, if there were a teller to tell them well.”

As caregiver members of the larger PH community, we are storytellers learning to tell those stories well!

The story that emerges for me about our caregivers is one of courage, caring, and compassion. Your answers show me a community that cares deeply for each other. You draw courage and strength from sharing your experiences, frustrations, tribulations, and triumphs with each other through Facebook groups, Meet-Ups, and your local support groups. You participate in fundraising and awareness activities in your neighbourhood and through national campaigns. You

manage medications and numerous appointments for your loved ones, often on top of full-time jobs, raising your family, and anything else that “normal life” demands of you.

Are you starting to see the “heroic” qualities of who you are and what you are doing?

Some of you started this journey many, many years ago (ranging from one year since diagnosis up to 25). Some of you have relocated to be closer to a PH clinic and have easier access to treatments. The challenges that you face as your loved one progresses along the road (changes in treatment protocol, adverse reactions to medications, multiple and increasing visits to ER) are numerous and often overwhelming. And yet, you persist!

If I had to choose one word to describe the caregivers in our community, it would be resilience!

Psychologists define resilience as the process of adapting well in the face of adversity, trauma, tragedy, threats, or significant sources of stress. They call it a life skill. I define it as the quality we most need to develop from the day our loved ones are diagnosed with any form of pulmonary hypertension! The keywords are “adapting well.” Like many caregivers, I know there were days (weeks, months even) on my journey with Harry that I did NOT adapt well. And yet, there were many other days where we found the grace to be in the flow of our journey with laughter, love, and hope.

92% of caregiver survey respondents agreed or strongly agreed that they had contributed to an improvement in their loved one’s quality of life (made life easier and more manageable). This is cause for celebration! In turn, PH physicians, nurses, and patient organizations show up as your most effective sources of emotional support.

## From the 2021 Canadian PH Community Survey

82% of caregivers were married or common law  
6% were in a relationship but living separately  
11% were single

44% were the primary caregiver a spouse/partner diagnosed with PH  
30% were the parent/guardian of a person diagnosed with PH  
12% were the primary caregiver of a parent diagnosed with PH  
10% were the primary caregiver of another family member  
4% were the primary caregiver of a friend/colleague  
29% were also the caregiver of others as well as someone with PH

The survey also showed us what you need as the journey continues. When asked to rate the quality of your connection to your peers in the PH community, 46% rated it as excellent or good, but 24% found it only to be fair, and 31% felt it to be poor or very poor. Finding a way to strengthen connections in the caregiver community will be a crucial element in keeping you strong and resilient.

92% of caregiver survey respondents agreed or strongly agreed that they had contributed to an improvement in their loved one's quality of life (made life easier and more manageable). This is cause for celebration!

Having a peer group that shares the journey with you is so important. Knowing that you are not alone in what you are feeling or experiencing, learning from others who have been on the road longer, and leaning into a support group are essential. Some are formal support groups; others are informal networks such as those found through social media platforms. If you cannot find a group that matches what you need emotionally, then it is important you reach out to PHA Canada and ask for what you need.

The emotional roller coaster of the caregiver's life is sometimes very hard to share with those we love. 54% of you are comfortable letting your loved ones know how you feel, but there is 20% that disagree. One of the reasons I was so keen to create support group sessions for "caregivers only" was so we could express our concerns, fears, frustrations, and sometimes anger in a safe place. Bend before you break or vent before you burn out are essential elements to developing and maintaining resilience.

Coping skills and future treatment options showed up as the top two types of information you want for your loved ones and for yourselves. 85% said we need to increase the level of understanding of the public about the disease itself, and 98% said public awareness is a real issue. Almost half of you felt isolated

and excluded from society because PH is not a "visible" disease and acknowledged that you needed access to counselling/emotional support.

Your voices are being heard! Your story is being read and shared. And solutions, tools, and resources to help you where you need it most are being developed.

From my perspective, as one who has travelled the entire PH road with my loved one, I find your story, as written in these survey answers, to be one of hope, courage, and incredible resilience. I hope you see how strong, brave, and resilient you are. Mostly, I hope you know that no one expects you to write your story on your own. Communicating in community and sharing our stories are the most powerful tools we have to survive, so we can help our loved ones thrive.

Contributed by: Teri Kingston, Saint Andrews, New Brunswick

# The Canadian Pulmonary Hypertension Registry (CPHR): An Update



One of the more exciting and significant efforts undertaken by the PH medical community in Canada in the last five years is the Canadian Pulmonary Hypertension Registry (CPHR). CPHR is a database where anonymized information about pulmonary hypertension patients is stored. The data is used to understand how pulmonary hypertension affects Canadians compared to other countries to improve patients' care.

In the recently completed Canadian *PH Community Survey*, participants were asked a series of questions regarding the registry. Only 22% of respondents said that their, or their loved one's, medical information was included in the registry. Most respondents (73%) were uncertain if their data was included but were willing to provide blood and/or tissue samples for storage in the registry. Very few respondents had concerns about being a part of registry. Those who did have concerns reported that it was due to lack of knowledge about the registry or concerns about the security and privacy of their information.

It is evident by the results of the survey that most patients are interested and have very little reservations about participating in the registry however, most are not participating and are simply unaware of the program and its benefits.

So, to encourage more patients to participate, PHA Canada asked some of the key people behind the development and maintenance of the registry to tell us more about the registry and how it works.

## About the Registry

CPHR was created in 2017 to understand pulmonary hypertension in the Canadian context. The initial site was in Vancouver. In the first year, both Hamilton and Calgary joined CPHR and started entering data about their patients. Ottawa joined in 2018 and Halifax in 2019. Moncton, Winnipeg (SBGH), Québec City, St. John's, Edmonton, and Toronto have recently joined. Other centers (London, Kingston, and two pediatric centers) have been taking steps to include their patient population in the registry.

In total, 866 PAH patients and 238 CTEPH patients have been entered into the registry. A variety of information is collected during patient visits, including data from echocardiograms, blood work, right heart catheterizations, and six-minute walk tests. Oxygen and medication use are recorded if a patient has gone for a pulmonary endarterectomy or a transplant. If a patient in the registry passes away, information is sought to determine whether a patient died from pulmonary hypertension or other causes.

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There is a lot of change occurring within the rare disease drug development and approval process both nationally and globally. Patient registries and biobanks (repositories of blood samples) will play a key part in many of these changes.

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## Safety and Confidentiality

The CPHR data is stored on a secure server at the University of British Columbia. Data is entered into the registry by your health care providers or research assistants. When the data is pulled from the registry to be studied, it is anonymized and aggregated, meaning a patient's name is not associated with any data, and individual data is examined as part of a group. A research ethics board must review and approve any data access for studies.

If you are a patient in one of the centers listed above, don't hesitate to ask your health care provider if your data is being used in this important initiative and any other questions you may have about the registry.

## Research Publications to Date

The CPHR allows researchers across Canada to access data from the registry in a safe way that preserves confidentiality. Several research studies using the data have already been published. One of the unique characteristics of our country is its large geographic size and relatively small population. In a recent study, Dr. Nathan Brunner looked at patients who live in smaller communities who must travel long distances to be seen in the Vancouver PH Clinic<sup>1</sup>. The study results showed no association between community size and time to travel to the clinic with the severity of illness at diagnosis or prognosis at follow-up. This suggests that PH patients in remote communities in BC are receiving care for their pulmonary hypertension comparable to those near Vancouver's clinic.

An important study led by Dr. Lisa Mielniczuk in Ottawa was recently published that looks at the survival of PH patients over time in Canada<sup>2</sup>. The 1, 3, and 5-year survival rates in the Canadian population are comparative to populations in other large registries in the United States and Europe. The study also showed that Canadian physicians shifted towards using multiple pulmonary



hypertension medications earlier in the course of disease following the publication of the most recent ERS guidelines, which recommended that treatment plan. The treatment plan did not appear to improve survival in Canadian patients compared to earlier treatment strategies.

Other studies published have looked at the treatment of patients with pulmonary hypertension and left-sided heart disease as a comorbidity<sup>3</sup>.

Upcoming studies include looking at outcomes of patients who have transitioned from IV therapy to oral therapy, patients who have had a pulmonary endarterectomy in Toronto, and the quality-of-life assessment tools. An important future study by Dr. Brunner and Dr. Jason Weatherald (from Calgary/Edmonton), looking at outcomes in the Indigenous Canadian population, was also recently proposed.

## And That is Just the Beginning

There is a lot of change occurring within the rare disease drug development and approval process both nationally and globally. Patient registries and biobanks (repositories of blood samples) will play a key part in many of these changes. Aggregated and

anonymized patient data and technological advances are providing researchers with a treasure trove of data to discover previously unknown similarities (i.e., genetic mutations) and potential new treatment pathways among patients within a disease population. Drug manufacturers use this information to develop new and innovative therapies to improve the length and quality of patient lives. Sponsors of clinical trials are working with drug approval agencies to adapt clinical trials to better fit the needs of small, rare disease populations. Part of these adaptations will mean the inclusion of patient-generated information on the quality-of-life issues, alongside clinically relevant milestones. Drug approval agencies are also looking at how to get new therapies to a greater number of rare disease patients quickly by trying to determine what and how patient-reported information can improve and speed up their decision-making. What is common across the spectrum of drug development and approval is that patient registries will be an important source of the patient-generated information used throughout the process.

Contributed by: Joan Paulin, PH Caregiver, Mississauga, ON; Lisa Kolkman, Nurse Practitioner, Vancouver General Hospital, Vancouver, BC; Lena Legkaia, CPHR Project Manager, Vancouver, BC

## Building a National Strategy for Drugs for Rare Diseases

What We Heard from Canadians



Canada

For further information on how rare disease patient voices and data are having an impact at the regulatory level, please visit “Building a National Strategy for Drugs for Rare Diseases: What We Heard from Canadians” at [www.Canada.ca](http://www.Canada.ca). It’s quite long, but the executive summary is worth the read.

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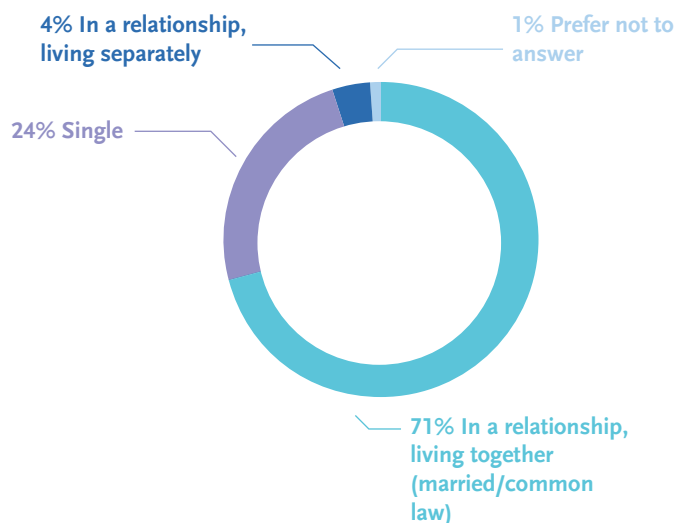
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# Demographics Report: A Snapshot of Who Responded to the Canadian PH Community Survey

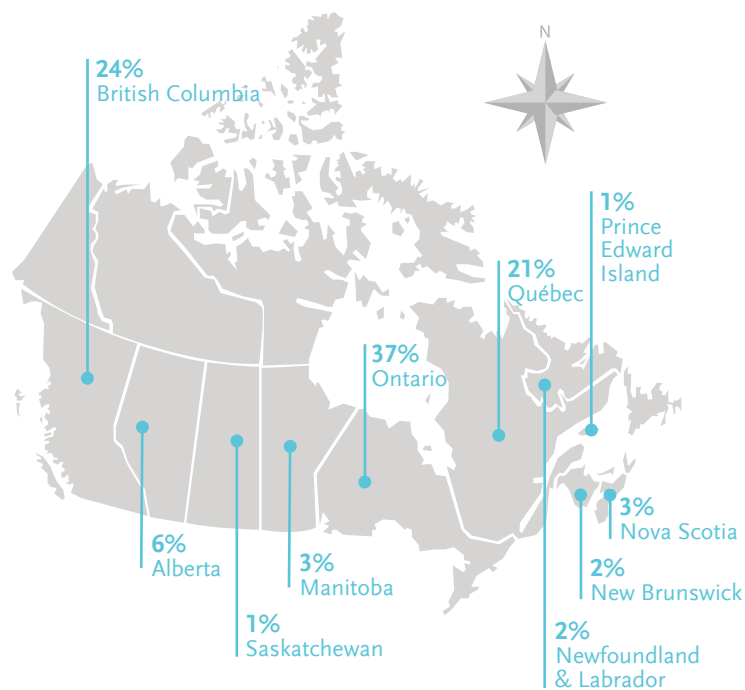
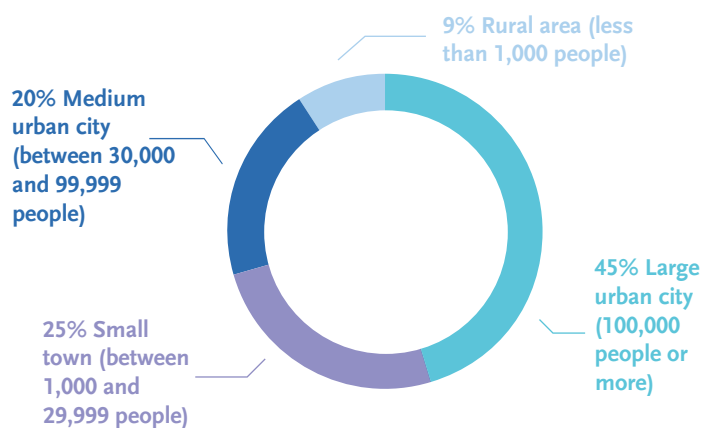
## Age/Gender

	Patients	Caregivers
Mean age	58 years old	53 years old
Male	11%	41%
Female	88%	59%
Non-binary/ third gender	0.3%	—

## Relationship Status



## Location



# Research Corner

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Since 2016, PHA Canada has awarded \$120,000 in scholarships to 11 talented researchers through its PH Research Scholarship Program. In this section we introduce you to our 2021 scholarship recipients and provide a fascinating research update from 2018 recipient Dr. Francois Potus about his investigations into the role of epigenetics in the development of PAH.

# Scholarship Recipients

Research brings hope. It also holds the key to a better understanding of pulmonary hypertension, leading to the development of novel therapies, improved treatments, and more sophisticated tools to care for patients. Research also holds the promise of a healthier and longer life, better management of symptoms, and ultimately a cure. PHA Canada is proud to help build capacity within the research community by awarding \$10,000 scholarships to outstanding trainees in support of their research in the field of pulmonary hypertension. We are pleased to announce PHA Canada's 2021 PH Research Scholarship Recipients Yann Grobs and Pierce Colpman.

## Yann Grobs



Yann Grobs, PhD student, Pulmonary Hypertension and Vascular Biology Research Group, Laval University, Quebec City, QC

### Scholarship

The Loretta Chu Memorial PH Research Scholarship was awarded in 2021 to honour Loretta Chu and her many contributions to the PH community, including during her many years as the co-leader of the Toronto support group. This special memorial scholarship was funded thanks to a major donation by PHA Canada Founder and Eternal PHriend, Darren Bell, and the fundraising efforts of the PH community during *GolPH for PH* in June 2021 (see page 11).

### Recipient

Native from Montpellier, France, Yann Grobs obtained a bachelor's degree in health biology before specializing in microbiology and immunology during his master's degree at the University of Montpellier II. During an internship at the Research and Development Institute (IRD) on the genetic diversity of HIV, Yann developed a particular interest in translational research and, more particularly, in the physiology of the cardiovascular and respiratory

systems. Pulmonary arterial hypertension (PAH) being one of the complications of an HIV infection, Yann enthusiastically joined the PAH group of Québec, led by Professors Sébastien Bonnet and Steeve Provencher, to carry out a PhD under the supervision of Dr. Bonnet. After studying the involvement of the FOXO3 transcription factor in the development of PAH, Yann continued his work in epigenetic modification and focused his interest on the impact of histone acetyltransferase (P300) in PAH development.

### Yann's Project

*P300/CBP: A potential therapeutic target to cure pulmonary arterial hypertension*

### Project Description

Pulmonary arterial hypertension (PAH) is a progressive and incurable vascular disease. This disease affects the pulmonary arteries (PA) that carry the blood from the heart to the lung. PA are crucial for the reoxygenation of the blood. Like in cancer, cells in the walls of PA multiply in an abnormal manner (i.e., faster than normal) and lead to obstruction of the PA. This obstruction increases pressure in this vessel (the PA) leading to cardiac dysfunction. We identified P300 as a factor implicated in this gene expression. In cancer, its overexpression induces an increase in cell proliferation.

Similarly, we believe that this same factor is involved in abnormal PAH cell proliferation. We demonstrated that P300 is increased in PAH patients compared to non-PAH patients. In a PAH rat model, we showed that P300 treatment decreases PA cell proliferation and improves survival. We propose further investigating P300 function in general, which will benefit several diseases and improve the understanding of the development of PAH. Interestingly, a P300 treatment is currently in clinical trials for the treatment of cancer. Therefore, our study has high translational potential, including the development of a novel treatment strategy to improve PAH patient quality of life right from the laboratory.

# Pierce Colpman



Pierce Colpman, MSc candidate, Department of Medicine, Queens University, Kingston, ON

## Scholarship

The Mohammed Family PH Research Scholarship was founded in 2017, thanks to funds raised by siblings Renae and Joseph (Ajax, ON), in honour of their mom, Judy Mohammed. The Mohammed Family's popular annual *Ajax Walk/Run for PH Research* has since raised over \$40,000 for this scholarship.

## Recipient

Pierce Colpman is an outgoing and adventurous student, born and raised in Victoria, British Columbia. Pierce moved from Victoria to Kingston, Ontario to begin his undergraduate career in 2017, and since then has completed a specialized bachelor's degree in life sciences. In the Fall of 2021, Pierce was accepted into the highly competitive Translational Medicine master's program at Queen's University, where he began the next chapter of his career. Pierce also has experience volunteering in clinical settings as an Emergency Department volunteer with the Royal Jubilee Hospital in Victoria and with the

Campus Observation Room in consultation with toxicologists from Hotel Dieu Hospital in Kingston. Pierce's research aims to reveal the role of "dynamin 2" as a key protein involved in the final step of mitochondrial fission, and hence determine how dynamin 2 impacts the progression of disease in pulmonary arterial hypertension. This research will also explore an entirely new way of inhibiting mitochondrial fission through the inhibition of dynamin 2 in the hopes of discovering new potential therapeutic targets for PAH.

## Pierce's Project

*Increased expression of dynamin 2, a key regulatory protein in the final stages of mitochondrial fission, promotes human and experimental pulmonary arterial hypertension*

## Project Description

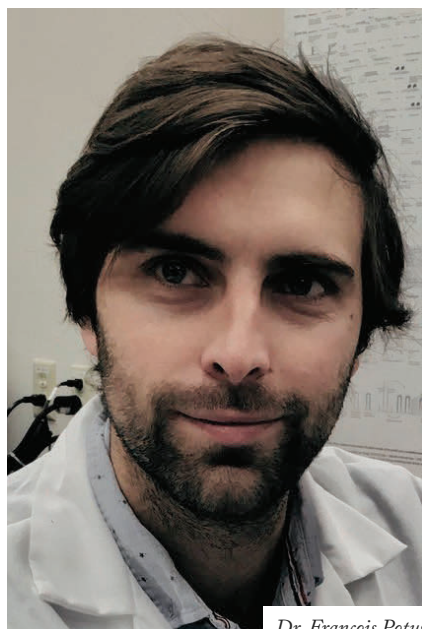
Many key features of pulmonary arterial hypertension (PAH), such as rapid cell growth of pulmonary smooth muscle cells and lack of programmed cell death (apoptosis), result from abnormal mitochondrial function. Mitochondria continuously join (fusion) and divide (fission) in coordination with the division of the cell's nucleus through mitotic fission, a very rapid process in PAH. We discovered that blocking mitochondrial division slows the growth of rapidly dividing PAH pulmonary arterial smooth muscle cells and can even kill them in experimental animal models of PAH.

Mitochondrial fission is controlled by a protein named dynamin-related protein 1 (Drp1); however, the final step in mitochondrial fission requires assistance from another protein: dynamin 2 (DNM2). DNM2 are expressed in excess in PAH patients, and therefore, we can block mitochondrial fission and slow down the growth of the cells by inhibiting DNM2.

This project will test the idea that increased DNM2 drives rapid cell division and that inhibiting DNM2 is a potential new form of PAH therapy. We will also study how excess DNM2 in PAH cells occurs by evaluating the microRNA that regulates the level of DNM2 as a potential diagnostic and prognostic biomarker to reveal new ways of treating and diagnosing PAH.

# The Role of Epigenetics in PAH: A Research Update

Francois Potus is a recently appointed Adjunct Professor at Laval University (Québec City, QC) and a young investigator in the Pulmonary Hypertension Research Group at the Québec Heart and Lung Institute. In 2018, he was awarded a PHA Canada Paroian Family PH Research Scholarship for his work on the relationship between inflammation and the progression of pulmonary arterial hypertension. Dr. Potus' project *Reprogramming gene expression as a driver of right ventricular dysfunction in pulmonary hypertension*, is investigating the role of a critical regulator of this epigenetic mechanism—called ten-eleven-translocase (TET2)—in the development of PAH. We are pleased to share with you an update from Dr. Potus on his work.



Dr. Francois Potus

## Reprogramming gene expression as a driver of right ventricular dysfunction in pulmonary hypertension

Although the original pathophysiological mechanism of pulmonary arterial hypertension (PAH) is localized within the pulmonary circulation, the severity of symptoms and survival is primarily associated with the ability of the right ventricle to maintain its function in the face of increased afterload. Right ventricular failure remains the primary cause of morbidity and mortality in patients with PAH. *Thus, the characterisation of the mechanisms associated with right ventricular failure is one of the priorities of contemporary research in PAH.* Functionally, right ventricular adaptation to increased afterload (pressure in the pulmonary artery) goes through a transition from **adaptive** (characterized by preservation of normal cardiac output, right ventricular ejection fraction, and exercise capacity, concentric right ventricular hypertrophy with minimal right ventricular dilatation, and fibrosis) to **maladaptive** (low cardiac output, reduced exercise capacity, myocardial apoptosis, fibrosis, progressive dilatation, decreased RV capillary density, and remodelling) phases before finally culminating to the development of right ventricular failure. However, there is tremendous heterogeneity in the right ventricular response to chronic pressure overload, with respect to the degree of cardiac hypertrophy, its effects on cardiac output, and the likelihood of progressing to overt right ventricular failure. Interestingly, the heterogeneity in the right ventricular response to increased afterload is not explained by differences in right ventricular mass or pulmonary artery pressure (afterload), suggesting other mechanisms may be involved. *However, the nature of these mechanisms is currently poorly characterized.*

Right ventricular failure is associated with multifactorial mechanisms. Mounting evidence indicates that several molecular pathways orchestrate the detrimental evolution of right ventricular remodelling. After a cardiac injury due to pressure overload, inflammation is sustained through the upregulation of cytokine release, leading to fibroblast proliferation/activation, excessive extracellular matrix production, and thus impaired relaxation and contractility. Furthermore, oxidative stress and associated DNA damage response and a metabolic remodelling trigger the hypertrophic and profibrotic signalling cascades, resulting in cell death and progressive cardiomyocyte loss. *An efficient treatment for right ventricular failure in PAH should consider and target all these dysfunctions.* Several studies demonstrated that right ventricular failure is associated with impaired gene expression. In other words, the way that genes are expressed is different in failing and normal right ventricles. For example, we and others reported that over 5,000 genes are differentially expressed in failing right ventricles compared to normal right ventricles. *However, the mechanism(s) leading to adverse regulation of gene expression in PAH failing right ventricle remains, so far, unknown.*

## Epigenetics, DNA Methylation & Right Ventricular Failure

Our previous work pinpoints the role of “epigenetics” in the adverse regulation of gene expression in PAH etiology and RV dysfunction. Epigenetics is defined as a “stably heritable phenotype resulting from changes in a chromosome without alterations in the DNA sequence” and includes mechanisms such as histone modifications (e.g., acetylation) and DNA methylation. DNA methylation is a mechanism by which cells or organisms adapt gene expression in response to their environment, including pathological or physiologic stressors, such as inflammation, metabolic changes, and hypoxia. It is characterized by the addition of methyl groups on DNA nucleotides that impairs how genes are expressed. DNA methylation is a dynamic process that reflects the balance between the activity of DNMT (writers that add methyl groups) and TET enzymes (erasers that remove methyl groups). Thanks to the PHA Canada Paroian Family PH Research Scholarship, we recently identified a novel deleterious mutation in a member of the TET family (TET2) associated with a ubiquitous decrease of the TET2 expression in the peripheral blood cells of PAH patients.

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Similarly, impaired expression of a gene coding for a member of the DNMT family (DNMT3B) contributes to the development of PAH. Taken together these observations suggest that the DNA methylation landscape is impaired in PAH. Supporting this observation, we demonstrated that right ventricular fibroblasts extracted from a preclinical model of PAH with right ventricular failure are hyper-methylated and maintain a pro-proliferative and pro-collagenic phenotype in-vitro. Finally, we reported that increased DNA methylation is associated with angiogenic defects contributing to right ventricular failure in PAH patients. *Nonetheless, the DNA*

*methylation changes associated with right ventricular failure and their functional impact on gene expression have never been characterized.*

I am pursuing the hypothesis that DNA methylation is a critical determinant of right ventricular failure in PAH. In other words, we hypothesize that changes in DNA methylation could reprogram gene expression and lead to right ventricular failure. Consequently, targeting DNA methylation could reverse pathological gene expression and slow down or prevent the development of right ventricular failure. We are investigating the ability of epigenetic drugs that modify DNA methylation to prevent right ventricular dysfunction. Furthermore, the characterization of DNA methylation changes involved in the transition from normal toward compensated and decompensated could lead to identifying a novel class of biomarkers. We are working on the identification of DNA methylation changes associated with right ventricular dysfunction and survival in PAH.

With each new wave of understanding comes new opportunities for the rational design of novel therapies and biomarkers. Characterization of DNA methylation changes associated with adverse gene expression in failing right ventricle might open the door to the identification of a novel class of biomarker and potential therapeutic target in PAH right ventricular dysfunction.

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Contributed by: Dr. Francois Potus, Adjunct Professor, CRIUCPQ, Laval University, Québec City, QC

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