PULMONARY HYPERTENSION
ASSOCIATION OF CANADA

L'ASSOCIATION D'HYPERTENSION
PULMONAIRE DU CANADA

CONNECTIONS

The Official Magazine of the Canadian PH Community

Special Feature:

Work & PH

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Editor in Chief
Jamie Myrah, Executive Director

Contributing Editor
Kimberly Brunelle, Director, Strategic Initiatives

Contributing Proofreader
Kimberly Brunelle, Director, Strategic Initiatives

Translation
Jeremy Durand, Manager, Marketing and Communications

Art & Design
Jeremy Durand, Manager, Marketing and Communications

Printing
BP Media

Contributors
David Christiansen
Nicole Dempsey
Don Downey
Kathy Downey
Krista Kemp
Nicole Klassen
Saeid Maghsoudi
Sanjay Mehta
Jamie Myrah
Mahnaz Nazari
Ashok Pandey
Brooke Paulin
Joan Paulin
Kaitlyn Salonga
Mitesh Thakrar
Rhea Varughese

In This Issue

Inside PHA Canada

Your Community

Work & PH Feature

Research Corner

Message from the PHA Canada Team

Empowering Lives: From Diagnosis to Daily Living with PH

Hello PHriends,

As we say goodbye to 2024 and welcome another new year, we are grateful to bring you the latest issue of Connections Magazine. In this issue, we will delve into some of the realities of the impact of pulmonary hypertension (PH) on the work lives of patients and caregivers. We also bring you updates from some of the critical projects we've been working on, including World PH Day, our annual PH Community Conference, and the launch of our sotatercept advocacy campaign, Time Matters PAH. Our team has been hard at work educating and supporting the PH community, spreading PH awareness, and creating opportunities for you to share your stories and contribute to better PH care and treatment. As we turn the calendar to 2025, it has been a time of reflection and planning for us at PHA Canada. We have paused to rest and connect with our loved ones, and we hope you have, too! We have also taken time to appreciate the many people who have contributed to our successes over the past year and to prepare for future changes and opportunities. Change is a natural part of the season, and it's also a part of our journey as an organization.

We are excited to share that we have welcomed a new Board Chair, Joan Paulin, to lead us into the next chapter of our work. Joan took over the role from Dr. Lisa Mielniczuk who recently left her position at the Ottawa Heart Institute and moved her PH practice and research to the Mayo Clinic in Minnesota, USA. We are so grateful to Lisa for her leadership and continued service to the community as a Board Director and member of the Canadian PH Medical Committee at PHA Canada. We wish her the best in her new role and look forward to collaborating internationally with her! We are thrilled to see Joan step up as Board Chair. Joan has been a passionate and hardworking advocate for the PH community since her daughter, Brooke, was diagnosed with pulmonary arterial hypertension in 2014. She brings valuable expertise in patient advocacy and the Canadian healthcare landscape. We know she will be an outstanding champion for PH patients as Board Chair. We are also pleased to welcome Sonya Collins (Paradise, NL) and Dr. Mitesh Thakrar (Calgary, AB) to the Board of Directors. Check out the Memo section to learn about our newest Directors and the latest changes at PHA Canada as our team continues to grow and evolve.

Change is something that the PH community knows all too well. Living with pulmonary hypertension means becoming accustomed to change, changes to how you feel, how you function, and how you manage your condition. Life can shift dramatically after a PH diagnosis, and so it's no surprise that PH can have a profound impact on work and career. It is common for people to face the difficult decision to reduce hours, change roles, or even leave the workforce altogether due to the physical and emotional toll of living with PH. In 2023, PHA Canada, in partnership with the Canadian VIGOUR Centre, a research group at the University of Alberta, conducted a study to help us better understand the socioeconomic impacts of pulmonary arterial hypertension (PAH) in Canada. We surveyed over 200 patients to learn how PAH impacts employment status, productivity and activity levels, finances, and quality of life. The survey also looked at similar impacts on caregivers.

In this issue's special feature section--Work and PH--you will get a glimpse of the survey data and the real-life stories behind what the numbers reveal. Our community contributors share their personal stories on navigating work demands, whether managing a career, balancing responsibilities, or considering alternative paths. These stories highlight that work is much more than earning a living. It's a source of identity, purpose, and connection. For families living with PH, finding a balance between managing health and maintaining this vital part of life is a journey of its own and one that deserves recognition and support.

In the Research Corner, we continue to explore the

Staff and Board, Vancouver, BC, November 2023

impact of a PAH diagnosis on employment, work productivity, and quality of life. Here, we highlight the considerable challenges PAH patients face in the workplace and how maintaining employment could be an important treatment goal for enhancing PAH patients' quality of life. In this section, you will also meet Mahnaz Nazari, the inaugural recipient of the PHA Canada Bell Family Pediatric PH Research Scholarship. You will receive an update on her groundbreaking research to develop cell-based therapies for premature infants suffering from PH.

While we recognize the benefits to patients and their caregivers of remain in the workforce, we know that isn't always possible for everyone. Without access to better treatments for all types of PH, patients and their caregivers will continue to experience significant barriers to participating in the workforce. PH patients need access to treatments that will not only slow or even stop the progression of their disease but also restore their quality of life and reduce their symptoms. This is one of the reasons we were pleased to see sotatercept (Winrevair) receive approval from Health Canada in August 2024 after a six-month priority review. Sotatercept (Winrevair) is the first activin signalling inhibitor for long-term pulmonary arterial hypertension (PAH) treatment. PHA Canada's Education Committee summarizes the findings from the phase 3 sotatercept STELLAR study in the Research Corner.

Since its approval by Health Canada in the summer, all eyes have been on sotatercept. This past fall, PHA Canada launched an advocacy campaign–Time Matters PAH–to help ensure this treatment becomes available to patients in Canada regardless of where they live or what type of insurance coverage they have. There are many ways for you to get involved! Join our electronic letter-writing campaign, contact your local government official to schedule a virtual or in-person meeting, and share messages across social media to help spread the word!

Our volunteers drive PHA Canada. We couldn't do what we do without them. Whether advocating to the government, planning the annual community conference, supporting newly diagnosed patients, writing articles for Connections, developing new resources, or raising crucial funds to support our work, volunteers are behind it all. Throughout this issue, you will see volunteers--leading as Directors and Ambassadors, advocating for change on Parliament Hill, supporting others by sharing their intimate experiences with PH, and so much more. Longtime volunteers like Jas James, who was recognized as an Eternal PHriend of PHA Canada at the 2024 Community Conference. New volunteers like Kaitlyn Salonga, Dr. Krista Kemp, and Dr. Rhea Varughese have brought critical new perspectives to our Education Committee. We want to take this opportunity to thank everyone who has supported us over the past year. If you haven't gotten involved yet, what are you waiting for? Whether you send a simple email to your local representative, become a volunteer, or donate to help expand and sustain our programs and services, you can make a difference today. Visit us anytime at www.phacanada.ca to learn how you can make 2025 a year to remember for all Canadians affected by PH.

Sincerely,

Team PHA Canada

Memo: Inside PHA Canada at a Glance

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

Welcome

Sonya Collins
Board Director, Paradise, NL

Diagnosed with IPAH in 2003, Sonya has been a passionate advocate for the PH community. She co-founded Atlantic Canada's PH Warriors and has served as a PHA Canada Ambassador since 2018, participating in committees, studies, and advocacy efforts. Sonya successfully advocated for financial support for PAH patients in her province, securing access to crucial treatments. Now, as a Board Director, she looks forward to continuing her work supporting the PH community.

Mitesh Thakrar
Board Director, Calgary, AB

Dr. Mitesh V. Thakrar is a pulmonary physician in Calgary, Alberta, and a Clinical Associate Professor of Medicine at the University of Calgary. He is the Division Head of Transplant Medicine and Medical Director of the Southern Alberta Solid Organ Transplant Program. After years as chair of PHA Canada's Education Committee, Dr. Thakrar now serves on the Board of Directors, bringing his expertise and passion for PH education to a broader leadership role. His research focuses on clinical outcomes in pulmonary hypertension, using the Canadian PH Registry.

Robyn Kalda
Manager, Research & Policy, Toronto, ON

Robyn recently joined PHA Canada as the Manager of Research and Policy. With a career in health promotion since the late 1990s, she has focused on maximizing the impact of information sharing, with special interests in plain language and the ethical use of technology. Robyn has worked with numerous equity-oriented groups at local, provincial, and national levels. At PHA Canada, she aims to enhance collaboration and information sharing between patients, researchers, healthcare providers, and policymakers to support the PH community. She lives in Toronto with her family and enjoys reading, cycling, and quilting in her spare time.

Goodbye

Joan Gibson
Ambassador, Ottawa, ON

We would like to share that Joan Gibson has retired from our Ambassador program. Joan joined the program after her daughter Jane was diagnosed with idiopathic pulmonary arterial hypertension in 2016. Joan has been a tireless advocate, using her professional expertise to champion better and more accessible treatments for those living with PH. Her unwavering support and dedication to this cause have left a lasting legacy. Thank you, Joan, for your years of service, your wisdom, and your heartfelt commitment to the PH community.

Pat Kelly
Project Manager, Research Capacity Building

We want to extend our heartfelt thanks to Pat Kelly, who recently completed her contract as Project Manager of Research Capacity Building. Over the past two years, Pat worked closely with our Ambassador team, helping them develop leadership skills and become patient partners in research projects like the upcoming CRAVE trial. She also played a crucial role in expanding the Canadian PH Registry and launching the Canadian PH Research Network, both key initiatives that integrate patient voices into the research process.

Sheri Elefant
Director, Fund Development, Westmount, QC

Sheri joined us in June as our Director of Fund Development, bringing her expertise and passion to advance our mission. While she has stepped back from her role for personal reasons, she has continued to support our fundraising efforts during this transition. We're deeply grateful for her dedication and look forward to welcoming a new Director in the new year.

Time Matters PAH:
Advocate for Sotatercept in Canada

#TimeMattersPAH

We're rallying the PAH community to urge provincial and territorial health leaders to prioritize and expedite sotatercept funding. PAH patients cannot afford to wait for essential treatment, and by joining this campaign, you can help expedite public funding for sotatercept, a new PAH treatment recently approved in Canada.

How You Can Get Involved

1. Write to Your Health Minister: Use our template letter to urge your provincial or territorial health minister to expedite funding negotiations for sotatercept. A few minutes of your time could make a real difference.

2. Share the Campaign: Encourage friends, family, and colleagues to join you in advocating for PAH patients by sharing our campaign toolkit and social media posts.

3. Stay Connected: Follow PHA Canada for campaign updates and additional advocacy opportunities to support timely access to new PAH treatments.

Your voice matters. By supporting Time Matters PAH, you're helping to secure faster access to life-saving therapies for Canadians with PAH.

Learn more about our Time Matters PAH campaign at
phacanada.ca/timematters

Your Community

Canada's PH community showcased incredible advocacy and awareness efforts for World PH Day 2024. We united to make a profound impact, from illuminating landmarks across the country in periwinkle purple for the Paint Canada Purple campaign to releasing significant research on the socio-economic burden of pulmonary arterial hypertension (PAH). On Parliament Hill, advocates met with elected officials to push for better support and access to life-changing treatments like sotatercept, with our mission highlighted during Question Period. The PH Community Conference brought patients, caregivers, and healthcare professionals together to share knowledge, foster connections, and discuss the future of PH care in Canada. These initiatives were fueled by the generosity of donors and fundraisers who continue to make life-changing work possible for the PH community.

Rare Disease Day

Rare Disease Day is observed worldwide on the last day of February to highlight the impact of rare diseases. We were honored to participate in the 2024 Rare Disease Day Summit! We were proudly represented by Ambassador Jennifer Bryson and Director Joan Paulin, who shared the voices of the PH community.

Learn more at phacanada.ca/rarediseaseday

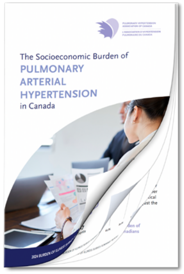
World PH Day

World PH Day 2024 marked an extraordinary effort by PHA Canada and the Canadian pulmonary hypertension (PH) community to raise awareness, advocate for better care, and celebrate the resilience of those affected by PH. This year's initiatives showcased a combination of public engagement, research dissemination, direct advocacy, and strategic media outreach, all aimed at making a significant impact on the lives of Canadians living with PH.

Learn more at phacanada.ca/wphd

The Paint Canada Purple Campaign

One of the highlights of World PH Day in Canada is the "Paint Canada Purple" campaign, which each year lights up landmarks in periwinkle purple on May 5th to honour those living with PH. This year, 23 monuments across the country were illuminated, and seven official proclamations recognized World PH Day. From coast to coast, the color purple brought visibility and hope to the PH community.



The Socioeconomic Burden of Pulmonary Arterial Hypertension (PAH)

In partnership with the Canadian VIGOUR Centre, PHA Canada released a report detailing the socioeconomic challenges of living with pulmonary arterial hypertension (PAH). The findings show the severe impacts of PAH on patients' financial stability and quality of life. This report aims to increase awareness and drive support for improved treatment options. Read the full report on PHA Canada's website.

Learn more at phacanada.ca/boi

Awareness Days on Parliament Hill

On May 6th and 7th, PHA Canada representatives gathered on Parliament Hill in Ottawa to advocate for enhanced support for PAH patients. Through personal stories and calls to action, they urged officials to prioritize rare disease funding under Canada's National Strategy for Rare Diseases. Their efforts were highlighted in the House of Commons by Marie-France Lalonde, MP, amplifying the need for action to support the PH community.

2024 PH Community Conference

The 2024 PH Community Conference in Calgary brought together Canadian PH patients, caregivers, and healthcare professionals for a weekend of learning, connection, and empowerment. Attendees engaged with inspiring keynote speakers, workshops, and panel discussions on the latest in PH research, treatment, and community support. If you missed the event, recordings of selected sessions are available on our website and YouTube channel at phacanada.ca/conference.






Eternal PHriends

The highlight of the opening reception was the recognition of Jas James as PHA Canada's newest "Eternal PHriend." Diagnosed with PAH 23 years ago, Jas has been a source of strength and hope for the PH community. From her work as a PHA Canada Ambassador to organizing support events and advocating for better care, Jas's unwavering dedication has made a lasting impact. Read more about this amazing individual and our other Eternal PHriends at phacanada.ca/EternalPHriends.



2023 Donor Recognition

We extend our heartfelt gratitude to everyone who supported PHA Canada last year. Whether as a monthly donor, major donor, event organizer, or sponsor, your contributions have made a tremendous impact. Your generosity has enabled us to provide vital resources, support services, and education to individuals and families affected by PH. Community events have strengthened our PH community, and sponsorships have backed key initiatives. Your commitment inspires our mission for a brighter future for Canadians living with PH. Thank you for making a lasting difference!

Major Donors

David Anthony
Darren Bell
Adam Blanchette
Jane Bondy
Ryan Brinkhurst
Dr. Nathan Brunner
Dr. George Chandy
Wing Chu
Barbara Clement
Michael Comeau
Bruce Danforth
Sheila Dempsey*
Roger Dowdall
Donna Downes
Ian & Joan Gibson*
Melvin Griffin
Patti Guminny
Kimarie Harris
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Carol Skippen
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Dr. Duncan Stewart
Peter Wade

Monthly/Quarterly/Annually

Jeff Becker
Nasreen Bhalloo
Dr. David Christiansen
Dianne Curle
Dominic Dedato
Nicole Dempsey
Katherine Huisman
Doris Issa
Jasbir James
Grace Libuna
Karen Mayner
Julie Mikuska
Renae Mohammed
Cynthia Neilson
Judith Pederson
Garth Robertson
Birdeen & Carl Selzer
Marilyn Stubberfield
Ashley Sudsbear
Joan Yearsley

Community Events & Campaigns

Bedard Family Event
Chicago Title Insurance
GolPH for PH (Milton, ON)
6-Minute Walk for Breath (National)
Paypal Giving Fund Canada (Facebook Fundraising)
Personal Fundraising Page - Jane Shirley
Personal Fundraising Page - Ryan Park

Corporate & Fondation Donors

Anonymous
BP Media
Dubwear Inc
HGA Service Inc
Skipton Solutions Inc.

Corporate Efforts

We are pleased to recognize corporate supporters at the following sponsorship levels, based on the total amount of funding committed during 2023.

Supporters - Less than $25,000
Bayer
Gossamer Bio
SRx Health Solution

Bronze - $25,000 - $49,999
Shoppers Drug Mart Specialty Health Network

Silver - $50,000 - $74,999
Merck

Gold - $75,000 - $99,999
United Therapeutics Corporation

Platinum - $100,000 and above
Janssen

also Monthly Donors

GolPH for PH 2024

Joan and Pat Paulin, organizers of GolPH for PH:

"Ten years ago, we embarked on a mission to raise awareness and funds for PH. Our journey began with a simple yet profound idea: to unite passionate individuals in a common cause. Year after year, our dedication and unwavering commitment have turned GolPH for PH into a cornerstone event for PHA Canada and the PH community.

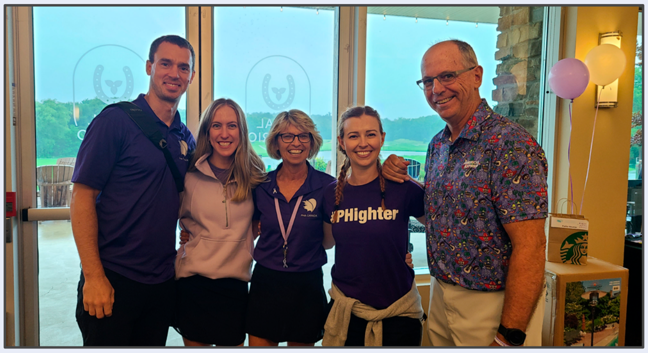
This year, as we celebrated the 10th annual GolPH for PH tournament, our hearts were filled with immense gratitude. The Paulin family's steadfast devotion has not only fueled our mission but inspired countless others.

With a fundraising goal of $30,000, we approached this year's event with high hopes and a determined spirit. Thanks to the incredible generosity of our supporters, we successfully reached this target. Each contribution is a testament to the community's commitment to making a difference.

It's crucial to recognize that 50% of every donation directly supported essential research. These funds help advance our understanding of PH, drive the discovery of new treatments, and improve the lives of those living with this condition. Your contributions are not just donations; they are investments in a future where PH patients can lead healthier, fuller lives.

The success of GolPH for PH wouldn't have been possible without the support of United Therapeutics Corporation, our generous sponsor, and the tireless efforts of our passionate volunteers. Your dedication turned this event into more than just a fundraiser; it became a celebration of community, resilience, and hope.

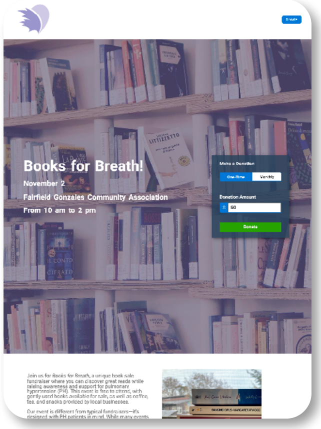
From the bottom of our hearts, thank you for your compassionate presence and the profound difference you make. Together, we are not just raising funds—we are lifting spirits, fostering hope, and changing lives. Let's continue to build on this incredible momentum and strive for a brighter future for all those affected by PH."



Celebrating Our 2024 Community Fundraisers

Dolores's Heavenly Heart

Dolores's Heavenly Heart hosted an unforgettable evening of stand-up comedy to raise awareness and funds for the PH community. By setting up their fundraising website through PHA Canada's platform, this event brought laughter and joy to attendees while supporting vital resources and awareness for PH. Thank you, Dolores's Heavenly Heart, for your dedication to making a difference for those living with PH.



Books for Breath!

November 2

Fairfield Gonzales Community Association

From 10 am to 2 pm

Join us for *Books for Breath*, a unique book sale fundraiser where you can discover great reads while raising awareness and support for pulmonary hypertension (PH). This event is free to attend, with gently used books available for sale, as well as coffee, tea, and snacks provided by local businesses.

Our event is different from typical fundraisers—it's designed with PH patients in mind. While many events

Books for Breath

Books for Breath, organized by Terri Gower, brought the community together for a one-of-a-kind book sale fundraiser. With gently used books, coffee, tea, and snacks for sale, this event raised funds and awareness for PH through PHA Canada's platform. By creating an inclusive and accessible space, the organizers helped support PH patients and ensure their voices were heard. Thank you for your creativity and commitment to the PH community!

Let's breathe together for a cause!

Our Friday evening Karma Classes this November are **donation-based**, and all funds raised will go to the **Breathing as One initiative** to support those living with **Pulmonary Hypertension**.

Book a class now at:
charlottetown.modoyoga.ca

Breathing as One Initiative

In November 2024, Modo Yoga Charlottetown made a meaningful contribution to PH Awareness Month by hosting donation-based Friday evening Karma Classes. These special classes invited the community to come together, raise funds, and support the Breathing as One initiative for those living with pulmonary hypertension. Thank you, Modo Yoga Charlottetown, for your generosity and commitment to raising awareness and making a difference!

Fundraising Made Easy

Create a Personal Campaign: Set up your online fundraising page and invite your network to support the cause. It's quick, easy, and impactful.

Host a Virtual Event: Bring people together for a fundraiser from the comfort of your home—trivia nights, bake sales, or anything you dream up!

Learn more at phacanada.ca/fundraise

Special Feature

Work & PH

Living with PH can make balancing work and daily life difficult. Symptoms like fatigue and breathlessness may limit your activities, and frequent medical appointments can mean taking time off. For caregivers, the challenge of managing their own work while supporting a loved one adds extra stress. These limitations can feel overwhelming, but understanding your options for workplace accommodations, support systems, and ways to manage daily tasks can make a difference. By addressing these challenges head-on, both patients and caregivers can find ways to adapt and maintain a sense of balance and control.

Balancing Work and PH: Our Journey Through Uncertainty

Kathy and Don Downey, from Calgary, Alberta, have been navigating the challenges of PH since Kathy's diagnosis on March 13, 2020—coinciding with the start of the COVID-19 pandemic.

Navigating the complexities of pulmonary hypertension and working can be daunting. Throw in a global pandemic, and it becomes even more complex. This was the situation we were flung into when Kathy received her pulmonary hypertension (PH) diagnosis on the same day that the world shut down for COVID-19 in March 2020. It was only appropriate that it was Friday the 13th.

Many factors are involved when dealing with work and PH. Family dynamics, work history, job availability, and balancing work with the realities and responsibilities of PH are some of the primary factors. These factors can be dramatically different for each PH patient and their family. We will cover our story and hopefully provide some insight into how we've dealt with working while being a household that includes a PH patient.

Kathy had been working at the local university but was one of the victims of mass layoffs due to government cutbacks in November 2019. After taking some time off for the Christmas holidays and a well-deserved vacation in San Diego that had been booked before she was laid off, she began searching for a new position in February 2020. The week after her diagnosis, she was put on high-flow supplementary oxygen. When things started opening up again after the initial COVID shutdowns, it was readily apparent that she wouldn't be able to work anymore. She applied and was accepted for provincial and federal disability benefits. This helps with our monthly expenses but never comes close to the income level she had been bringing in at the university.

Don had spent over a decade working from home as Director of Operations for a small niche publishing company specializing in forensic pathology. All the authors they worked with were full-time medical examiners writing reference books on top of their regular work. When COVID hit, their work caseloads went through the roof, and all of the projects the company was working on were put on indefinite hold, which is where they remain to this day.

The house we lived in with our two adult children was a two-storey with a walkout basement that had been converted into a separate suite where Kathy's parents lived. We had already taken on a role as caregivers for Kathy's parents, who were in their mid-to-late eighties when she received her diagnosis, and the logistics of her high-flow oxygen put the majority of those responsibilities on Don along with his new caregiver responsibilities brought on by Kathy's PH. Don started looking for work but couldn't find anything that fit his qualifications.

He decided to return to school in the fall of 2021 and take a two-year program in automotive management. In the summer between school years, our daughter took a summer job at a hotel in Waterton Lakes National Park, which is almost 300 km from Calgary. She told Don that the company was looking for additional staff, and he took a position at one of their other hotels. Don was quickly promoted to front desk supervisor and by the end of July, he had been asked to stay on for a year until the end of the next summer. After some discussion, we decided to go for it and arranged for Kathy to divide her time equally between Calgary and Waterton, switching every two weeks. By mid-October, Kathy was having issues with her PH, and the doctor said that it had to do with the frequent elevation changes as Waterton is 245 m (800 ft) higher than Calgary. He told her that she would have to decide on one location or the other, and we determined that it would be logistically better for her to be in Calgary. Don resigned from his position so we could be in one place. He could resume his studies in January and complete the third semester of his program.

By the late spring of 2023, Kathy was struggling to climb "Mount Everest," the stairs from the main floor to the upper

floor where our bedroom was. Meanwhile, her parents were dealing with mobility issues and decided that they should move to the seniors' lodge in our neighbourhood. We began the process of selling the house and preparing to move. Our son was moving to Edmonton, and we also decided to move there because the real estate market was considerably more affordable. We bought a condo apartment where we would be mortgage-free. Don started searching for work but had trouble finding anything that would allow him the flexibility to work and get Kathy to all of her medical appointments. In late March of 2024, with Kathy's needs increasing, we decided to sell the condo and move back to Calgary. Don has taken on the caregiver role for Kathy's parents again, as he provides them transportation to all of their medical appointments.

As her PH has progressed, the number of trips to the PH clinic, the number of tests that require scheduling and attending appointments, and the number of hospital admissions have all increased considerably. In 2022, Kathy was forced to give up driving, so Don is the primary means of transportation to and from these appointments. Finding work that pays well enough and provides the flexibility to take Kathy to appointments is key.

In Don's job search, we believe certain factors could have been hindering him. Don is in his late fifties, and when employers look at his resume, they may think he will only work for a short time before retiring. Unfortunately, this is not the case, and he will have to work for several more years. Another factor is that he is applying for positions that are lower paying than his previous employment, and we believe that employers may feel that he is overqualified and will end up leaving for something else at the first opportunity when he just wants to get a position with one company and stay there until he eventually retires. Adding these factors to the need for flexibility makes finding a new job difficult.

We have been fortunate enough to make it financially, with some timely loans from Kathy's parents when we needed them that have since been repaid, but this has depleted our savings.

Contributed by: Kathy Downey, Living with PAH since 2020, and her husband and caregiver, Don Downey, Calgary, AB.

How I've Made Working Work for Me

One of the most challenging aspects of seeking accommodations for chronic illness is that many of the things you need can seem like luxuries to other people.

Brooke Paulin, a dedicated professional from Mississauga, Ontario, has been living with PAH for over a decade while continuing to work full-time.

To briefly introduce myself, my name is Brooke Paulin, and I'm a thirty-something-year-old from Mississauga who was diagnosed with pulmonary arterial hypertension (PAH) over ten years ago and who continues to work full-time while managing PAH.

To say working with a chronic illness can be tough is an understatement–it's no secret that the world of work isn't yet particularly aware of, let alone accommodating of, most invisible illneses. So it's all about learning how to navigate a work/life/chronic illness balance and make the most of things.

Before my diagnosis, I was a competitive figure skater and have never not been on the go, so following my PAH diagnosis, I knew I would continue to work if my illness allowed me to; however, it's important to note that this is balanced with my quality of life (I am not choosing work over a balanced life).

As a disclaimer, I recognize that I have been fortunate in my career path post-diagnosis with the support and flexibility provided to me, but most patients have not been so fortunate and have had to face additional hardship.

Here are three key things that have helped me continue to work post-diagnosis:

1. Learning How to Manage My Illness

Of the items on this list, learning how to manage my illness has been by far the hardest and has morphed throughout my PAH journey.

Anyone with PAH knows that a 'one size fits all' approach is not compatible with our illness, so it's important to understand what works best for you while remaining compliant with your medical team's directives and prescribed medications.

As previously mentioned, how I manage my illness has morphed multiple times throughout the past 10+ years through various medication treatments; how I now manage on all oral medications is vastly different than how I managed while on subcutaneous Remodulin™ (or IV Remodulin).

While I was on oral medications since my diagnosis in 2014, I was transitioned to subcutaneous Remodulin™ in March 2020 (or what I refer to as 'when the world locked down thanks to COVID-19'). While this was a frightening time to transition to a new medication, in hindsight, it was a blessing in disguise: I was able to continue working full-time (remotely, as all physical offices closed) while learning how to manage all things subcutaneous. Similarly, nine months later, I was transitioned to IV Remodulin while the world was still on lockdown, which provided me with an opportunity to re-learn how to manage my illness (and more specifically, my new medications) from the comfort of my home while integrating my work responsibilities without the workplace setting.

To be transparent, I'm not confident I would have been able to continue working in the office five days a week while transitioning to subcutaneous Remodulin (and later IV Remodulin).

2. Finding the Right Accommodations

As PAH patients, our bodies don't work the way most do (duh!), so it's important to figure out what makes balancing work/life/chronic illness easier and what contributes to everyday difficulties. Some accommodations will help you, and some won't, and you won't necessarily know until you've tried them out.

One of the most challenging aspects of seeking accommodations for chronic illness is that many of the things you need can

seem like luxuries to other people. For example, sitting in a desk chair may cause muscle/joint fatigue, so you may request considerations for an ergonomic chair or a standing desk. Sometimes, it's as easy as adapting your work setup to better support the non-medication side effects of PAH. However, avoiding such 'obstacles' isn't always an option, which means finding other ways to balance work and PAH.

I'm fortunate that my job is primarily desk-based and offers a flexible working model where employees work partially in a physical workplace and partly remotely. Similar to my comment under 'Learning How to Manage My Illness,' I'm not confident I would have been able to continue working full-time if my job requirements were more physical in nature or in-office permanently.

3. Building a Support Structure at Work

I recognize this last item is not possible in all workplaces, but if you're able to and your workplace is supportive, finding support in key colleagues who recognize your work/life balance, including managing a chronic illness, is invaluable.

Ensuring I don't take advantage of my colleagues' trust, knowing they will understand if I need to disappear for a few hours (to combat medication side effects or for a doctor's appointment, for example), has made me feel more able to take time to rest when I need it—which has made it easier to get my work done when I'm feeling well and up to the task.

Remember, if you choose to continue working post-diagnosis, your work/life/chronic illness balance may continue to evolve as you move through your PAH journey. Like PAH, there is no one-size-fits-all approach to balancing the many components of living with a chronic illness. Some things to keep in mind:

- Working with a chronic illness can be tough, but workplace adjustments can lessen the load you must carry.
- Not specifically work-related, but the better we take care of ourselves and utilize the mobility aids that can help us, the better able we will be to balance the things we need to.
- Many of us have to find a careful balance between managing our growing career ambitions and our frustrating abundance of fatigue. This is OK!
- Don't burn yourself out and overexert yourself just to feel accomplished.
- As humans, it's in our nature to always search for the next thing. Grant yourself grace when navigating work and chronic illness.
- Our day-to-day working lives can feel like a marathon sometimes. Find ways to manage your illness that work for you.

And if you ever feel you need more support than your work colleagues can provide, don't hesitate to join a PH support group (either virtually, such as Canadian PHriends on Facebook, or in-person, should your city host one). While no two journeys are identical, you will find there's always support ready to be given—all you need to do is reach out!

Contributed by: Brooke Paulin, Living with PAH since 2013, Mississauga, ON.

PH Community Quotes

How has living with pulmonary hypertension (PH) impacted your ability to work and what adjustments have you made to balance your work, life, and illness?

When first learned I had PAH, I ended up taking a year and a half off work to learn how to manage and then because I wasn't sure if a lung transplant was right around the corner. Upon returning, I've had to work much harder than I used too not only because I find my body wears out faster than it used to but also I feel I need to work to prove to everyone I'm still able to do the job. Not only has PH made it physically harder to do the job, it has also presented challenges like where I store my medications and take my breaks (the break room is upstairs). The medications all present their own challenges.

Samantha Russell, living with PAH since 2022, Braeside ON

Before my husband was diagnosed, after he was diagnosed, and even after he passed, PH has had a negative impact and toll on my worklife. Being alone while driving to work was when the tears flowed and questions were asked (that no one could hear or answer).

I work in a fast paced environment. Even though I tried to put on a different face when I walked into the office, many mistakes were made. My mind was elsewhere. I was thinking of the dreaded disease and how it was affecting me and my family. Each day I get out of bed, put my feet on the floor, put one in front of the other, head to work, and hope today is going to be a better day.

Henrietta Billard, caregiver to her husband who passed in 2024, Isle aux Morts, NL

At 78, I was diagnosed with pulmonary hypertension and could barely function. My husband prepared for my funeral as I struggled to breathe and needed a walker. Without him, I'd likely be in a care home. Regaining strength has been slow, and I know that working wouldn't be possible even if I were younger. I can do light housework but still need afternoon naps and can't stand for long. Losing the ability to drive has been tough, but joining a knitting group and an exercise program has improved my mental health and stability.

Jacquie Lindgren, living with PAH since 2020, Saskatoon, SK

Living with a chronic illness like pulmonary hypertension (PH) can make everyday tasks feel overwhelming. Unpredictable health challenges force you to prioritize, adapt, and tackle life differently.

When I was diagnosed with PH, I was just days away from becoming a first-time mom—a full-time job in itself. The emotions were overwhelming, and the unknowns daunting, but I faced it all head-on. Traditional full-time work became impossible, yet I discovered fulfilling ways to stay productive.

I embraced volunteering, from coaching soccer and leading Girl Guides to organizing shoreline cleanups and supporting school programs. These activities allowed me to give back while managing my PH and thriving.

The key is balance. Some days demand rest, while others allow you to accomplish more. Celebrate what you can do, pace yourself, and prioritize your well-being. Every journey with PH is unique and beautiful in its own way.

Jennifer Bryson, living with PAH since 2012, York, PE

Being diagnosed with pulmonary hypertension turned my life upside down. I went from managing a busy gas bar to being hospitalized, and when I left, it was with an oxygen tank by my side. Working around fuel was no longer an option, and even standing for long periods became impossible. The job I once loved was suddenly out of reach, and I had to come to terms with a new reality.

Mike Tuxford, living with PAH since 2014, Lucan, ON

The most significant impact pulmonary hypertension has had on my work life is the persistent fatigue associated with the condition, which is further exacerbated by some of the medications used to treat it. This fatigue has affected both my paid and volunteer work. To manage this, I must carefully balance my activities with periods of rest. I have also taken medical leave from my job in the public school system to cope with these challenges.

Additionally, being diabetic compounds the issue, as chronic illness-related fatigue is particularly demanding. It often feels like when you're out of "spoons" (a metaphor for limited energy), you're not just out of spoons, you're out of options. As a result, you have to adjust and structure work and volunteer activities to accommodate these limitations.

Lori Bregoliss, living with PAH since 2020, Kamloops, BC

Navigating Disability and Income Loss After My PAH Diagnosis

Nicole Dempsey is a former elementary school teacher from Ontario who was diagnosed with pulmonary arterial hypertension (PAH) in 2013. Having spent 12 years passionately teaching grade 6, Nicole's diagnosis forced her to leave a career she loved.

I had been working as an elementary school teacher when I was diagnosed with pulmonary arterial hypertension (PAH) in 2013. I loved my job, earned a great salary, and happily taught grade 6 for 12 years. I distinctly remember during my first year of teaching, having to choose a long-term disability (LTD) insurance plan. There were various options, each with a different deduction from my pay. As a young teacher, I couldn't imagine ever needing LTD insurance and naively thought, "Who even needs LTD insurance?" I selected a plan with lower coverage, meaning I would receive only 45% of my salary if needed.

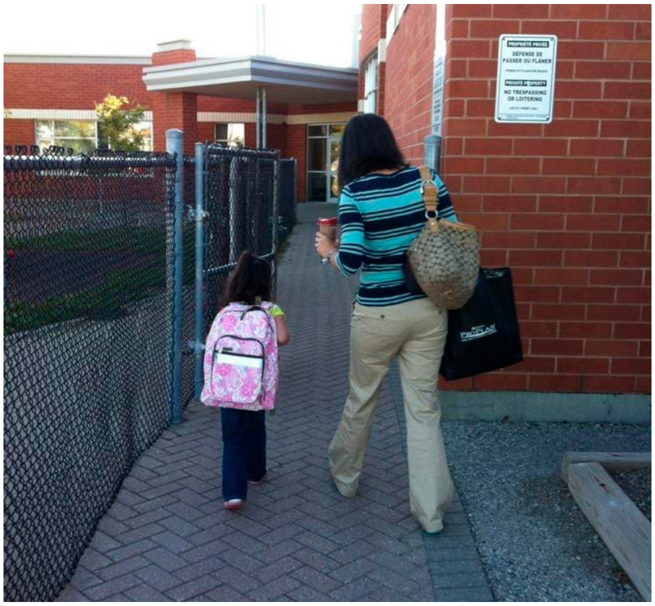
Unfortunately, things didn't turn out as I expected. When I received my diagnosis, I never imagined that I would be off work indefinitely. I held onto the hope that I might eventually return to teaching. However, as months passed, it became increasingly clear that returning to work wouldn't be possible. The progression of my disease required IV therapy, and the side effects made daily living extremely difficult. Some days were unbearable, and I couldn't envision standing all day while teaching young children.

After the first six months of being off work, my short-term disability insurance, which covered 100% of my salary, transitioned to LTD insurance at 45% of my salary. This substantial reduction had a significant impact on my family's combined income.

I never imagined not working. Growing up in a lower-income family, I had high hopes for my future. I attended university with the help of student loans, earning a Bachelor of Arts and a Bachelor of Education. I took pride in these accomplishments and in having a fulfilling and successful job. My strong work ethic meant that when I could no longer work, it took a toll on me mentally. I felt as though I had lost my sense of value and was no longer contributing financially as I had before my diagnosis.

I haven't worked in 10 years, and sometimes I feel embarrassed to admit this. The financial impact on our family has been significant, and the emotional toll has been equally challenging.

Contributed by Nicole Dempsey, Living with PAH since 2013, Cambridge, ON.



Juggling Career and Care:
The Reality of Parenting a Child with PH

Nicole Klassen is a dedicated mother navigating the challenges of raising a child with PH. After her son's diagnosis 12 years ago, Nicole took a career break to focus on his medical needs, balancing countless appointments, treatments, and unexpected illnesses alongside family responsibilities.

> On top of the typical time commitments that come with having children, we as parents to PH heroes now have both regular and unexpected medical appointments, medication changes or illness that can require a hospital stay.

It goes without saying that balancing work and family responsibilities is challenging for any parent. For those of us who have children with PH, we know that the experience can be even more complex. I had already anticipated that I would be taking a bit of a career break to stay home with our son but the plan was an eventual return to work. The reality is, most families need two incomes to make ends meet.

On top of the typical time commitments that come with having children, we as parents to PH heroes now have both regular and unexpected medical appointments, medication changes or illness that can require a hospital stay. Many times I have had to take days off or leave early if he's too unwell to attend school and no back up care is available. And understandably this can and has created tension with some employers and fellow employees as they count on their staff being present at their job, fulfilling their duties. We face the dilemma between being present at our job and fulfilling our child's medical needs, often feeling guilty about not being able to give 100% to either. With sporadic availability comes the possibility of not receiving hours or worse, termination of employment due to not being the right fit. I've experienced both of these and I am so thankful to now have two amazing employers and colleagues who have been flexible and supportive during our journey. Another big struggle is finding suitable child care for a chronically ill child, and in our case when he was younger, with subcutaneous IV infusion. When I first returned to work 10 years ago, many child care providers didn't have the ability, knowledge or willingness to learn to take on a child with extra needs. This still rings true today though we have been fortunate throughout the years to have had two wonderful dayhomes and a daycare who took him with no hesitation.

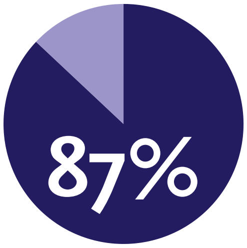
They say it takes a village to raise a child and I always say it takes an incredibly special village to raise a child with extra needs. After we received our diagnosis almost 12 years ago we started to build a network of supports in our community, some of which have become extended family and some have come and gone their separate ways. It's not perfect and it's not how we envisioned things but it works for us and hopefully in the future I will be able to return to working in a full time capacity.

The important thing is to remember that you are doing the best you can for your child or children and that is what matters. Keep building that village and let them help you. Even though it may seem impossible, take care of yourself . You can't pour from an empty cup.

Contributed by Nicole Klassen, mother and caregiver of a PH patient, Edmonton, AB.

The Socioeconomic Burden of Pulmonary Arterial Hypertension in Canada

In 2023, PHA Canada and the University of Alberta conducted a study to better understand the financial and employment impacts of PAH in Canada. The research aimed to understand how PAH affected patients' work and daily activities. Additionally, the survey captured data about patients' need for caregivers and the impact of supporting a patient with PAH on caregiver work productivity.



of patients reported that PAH limited their career options

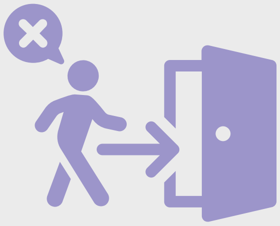
66%

of patients reported that PAH affected their ability to keep a job

61%

of PAH patients lowered their hours at work

Nearly 2 out of 4 PAH patients changed from full-time to part-time work



45%

of PAH patients reported having to quit working at their job

45% of patients indicated that due to their PAH, they currently require the help of a caregiver

Caregivers spent an average of **25 hours per week** helping with daily activities

Caregivers lost an average of **9 hours** per week of work time due to caring for their loved one with PAH

Learn more about the burden of PAH at
phacanada.ca/boi

Research Corner

This section highlights the latest findings from the phase 3 sotatercept clinical trial, the STELLAR study. Additionally, our Knowledge Philanthropy team provides insightful articles on how PAH impacts employment, work productivity, and quality of life. We're also thrilled to introduce Mahnaz Nazari, the first-ever recipient of the PHA Canada Bell Family Pediatric PH Research Scholarship. She will be sharing exciting updates on her pioneering research in pediatric pulmonary hypertension.

Sotatercept for the Treatment of Pulmonary Arterial Hypertension: The STELLAR Study

Sotatercept is a new molecule designed to work on the cellular pathways which cause blood vessels in the lungs to become constricted, overgrown, and narrowed. Small studies have shown that sotatercept can reverse some of these changes and open up these blood vessels. STELLAR was a **phase 3, double-blinded, randomized** clinical trial to confirm its effects. Its **primary outcome** was the impact of sotatercept on 6-minute walk distance (6MWD) in patients with pulmonary arterial hypertension (PAH). In PAH, 6MWD has often been used to obtain health authority approval of other medications for PAH.

A **phase 3** study is a large clinical trial comparing a treatment's safety and effectiveness against the current standard treatment. Phase 3 trials are done to try to receive regulatory approval for a medication to come to market.

A **double-blinded randomized** placebo-controlled trial is the gold standard of studies. Patients are randomly assigned to receive either the medication or a "dummy" medication (placebo). Neither the patient nor the investigator treating the patient knows which they are receiving (double-blinded).

A trial's **primary outcome** is the main question being asked. Secondary outcomes are other important measurements that are not the main criteria for success. A **hierarchical secondary outcome analysis** means that if the primary outcome is not positive, the second is not explored, and so on as one goes down the line.

In addition to 6MWD, STELLAR did a hierarchical secondary outcome analysis of multiple important secondary outcomes: multicomponent improvement, change in pulmonary vascular resistance, change in N-terminal pro–B-type natriuretic peptide blood level (NT-proBNP, a marker of how stretched out the heart is), improvement in WHO functional class (how short of breath someone feels), time to death or clinical worsening, French risk score, and changes in a patient's health-related quality of life (HRQoL), as measured by the Pulmonary Arterial Hypertension–Symptoms and Impact (PAH-SYMPACT) scores over 24 weeks.

Participants

STELLAR recruited 323 patients with PAH from 91 sites in 23 countries, including Canada. Participants were randomly allocated to receive either sotatercept (163 patients) or placebo (160 patients) via subcutaneous (under the skin) injection every 3 weeks for 24 weeks. All patients had WHO functional class II or III symptoms: they were symptomatic but not short of breath at rest. Patients in this trial were an average of 48 years old, with an average time since diagnosis of 8.8 years. All patients had already received approved PAH therapies for at least 90 days. Of importance, 61% were already receiving "triple therapy," and 40% were receiving an infusion prostanoid therapy. These two rates were much higher than in many previous trials of new medications. This allowed assessment of the potential benefits of sotatercept on top of many common therapies which patients may already be taking.

Results

The primary outcome was met. The minimally clinically important distance improvement (MCID)—the distance by which someone would notice an improvement in their

The authors of this trial concluded that sotatercept was an effective therapy for PAH, even when combined with currently available therapies.

symptoms—is believed to be 33 metres (m). People in the STELLAR sotatercept group walked, on average, 40.1 m further in 6 minutes, while the 6MWD for those in the placebo group declined by an average of 1.4 m.

Patients on sotatercept did better than placebo in all areas of HRQoL except for the Cognitive/Emotional Domains on the PAH-SYMPACT quality of life score. Sotatercept reduced the rate of clinical worsening by 84% over 32 weeks. Compared to placebo, it reduced mean (average) pulmonary artery pressure by 13.9 mmHg, which is a larger change than other PAH medications. Compared to only 10.1% in the placebo group, 38.9% of patients in the sotatercept group improved all three of 6MWD, NT-proBNP, and WHO functional class.

The authors of this trial concluded that sotatercept was an effective therapy for PAH, even when combined with currently available therapies.

Side effects

The placebo group had a higher rate of serious side effects (22.5% of participants) than the sotatercept group (14.1%). However, the sotatercept group had higher rates of bleeding (mostly non-serious, e.g. nosebleeds and bleeding gums), low platelets (cells which normally prevent bleeding), and higher blood hemoglobin levels.

Sotatercept is known to increase the risk of abnormal blood vessel growth called telangiectasia (pronounced tee-lan-jeck-tay-zya), which can lead to bleeding. Telangiectasia occurred in 10.4% of the sotatercept group versus 3.1% in the placebo group.

Limitations

- Relatively fewer patients with connective tissue disease PAH were enrolled than most clinics see, limiting the assessment of its effects and risks in patients with diseases like scleroderma.
- This trial did not examine the effectiveness of sotatercept in patients who are very early in their disease (WHO class I) or patients who are very ill with shortness of breath at rest (WHO class IV).

Next steps

The 40.1 m improvement in 6MWD and improvement in other markers of disease severity, despite many patients being on pre-existing therapies, is noteworthy. After this trial, sotatercept was licensed for use in the USA in March 2024, and on August 28th was approved by Health Canada.

Several ongoing studies will help us better understand the risks and benefits associated with the use of sotatercept in a broader range of patients with PAH.

Contributed by PHA Canada's Education Committee:
Dr. David Christiansen, Dr. Krista Kemp, Kaitlyn Salonga, Dr. Mitesh Thakrar, Dr. Rhea Varughese

Research Update: PAH Affects Employment and Patients' Quality of Life

PHA Canada Knowledge Philanthropists Saeid Maghsoudi and Ashok Pandey team up with PH specialist Dr. Sanjay Mehta to bring us two research updates about the effect of PAH on employment and patients' quality of life.

Impact of PAH on Employment, Work Productivity, and Quality of Life - Results of a Cross-Sectional Multi-Center Study

Jan Fuge, Da-Hee Park, Thomas von Lengerke, Manuel J. Richter, Henning Gall, Hossein A. Ghofrani, Jan C. Kamp, Marius M. Hoeper, and Karen M. Olsson. Frontiers in Psychiatry 2022; Vol 12: Pg 1-8.

Background. Research into the burden of a diagnosis of pulmonary arterial hypertension (PAH) on patients has focused on physical symptoms including shortness of breath and fatigue, reduced physical activity, and resulting poor quality of life. There is much less information on the effects of PAH on employment and work productivity, although a recent survey of 217 individuals by PHA Canada and the University of Alberta[1] found that 66% of patients with PAH reported that PAH affected their ability to keep a job, and those with jobs reported a 42% reduction in their work productivity. The current study surveyed patients in two large PH referral centers in Germany, assessing education, employment status, work productivity, and quality of life. Additional assessments of patients included clinical PH data, e.g., WHO functional class, 6-minute walk test distance (6MWD), and blood levels of NT-proBNP.

Findings. The study surveyed 212 adult patients with PAH including 72% females, with an average age of 57 years. Most subjects (76%) had idiopathic PAH, 10% had hereditary PAH, and 10% had associated PAH (e.g. connective tissue disease). Patients' educational backgrounds were categorized; most had a "moderate" level of education (63%; no university or college), with smaller numbers having "higher" (18%; university or college) or "lower" education (no formal education; 17%). At the time of PAH diagnosis, only 48% were employed (34% full-time, 14% part-time), which decreased significantly to 29% (21% full-time, 8% part-time) 6 years later at the time of the survey. The study identified this considerable loss of employment over the course of the disease, as well as a marked decline in work productivity as indicated by a Work Productivity and Activity Impairment score. Employed patients reported better health-related quality of life compared to unemployed patients.

Compared to patients with lower education, patients with moderate education were more than 3.6 times more likely to be employed, while those with higher education were 5.6 times more likely to be employed. In addition, the likelihood of being employed increased by 20% (odds ratio 1.2 fold) for every 50-metre greater 6MWD.

Implications. A diagnosis of PAH is associated with significant reductions in employment rates and work productivity over the course of the disease, and lower employment status is associated with worse quality of life. Patients with lower education levels and worse physical capacity (as reflected by shorter 6MWD) were less likely to be employed. The findings suggest that maintaining employment could be an important treatment goal for enhancing PAH patients' social participation and quality of life. The study underscores the need for targeted interventions to support employment among this patient group, potentially improving their overall well-being.

1. Pulmonary Hypertension Association of Canada. 2024. The Socioeconomic Burden of Pulmonary Arterial Hypertension in Canada Retrieved from https://www.phacanada.ca/boi

Social determinants of health in pulmonary arterial hypertension patients in the United States: Clinician perspective and health policy implications.

Vijay R. Nadipelli, Jean M. Elwing, Willie H. Oglesby, and Karim El Kersh. Pulmonary Circulation 2022; Vol 12, Issue 3: Pg 1-14

Background. Pulmonary arterial hypertension (PAH) is a complex, chronic disease that profoundly impacts a person's physical and mental health. Nonmedical social determinants of health (SDH), including an individual's socioeconomic status, housing, education, neighborhood, employment, social support, access to nutritious food, safe living environment, and access to healthcare can also contribute to poor health outcomes in many illnesses. These authors surveyed PH physicians at 17 US PH centres and 1 PHA US patient advocate to assess awareness of SDH principles and the impacts on people with PAH.

Findings. Although SDH information was not discussed or routinely collected at the PH centers, physicians surveyed had a good understanding of SDH principles, and the interconnectivity between an individual's health and factors such as healthcare access, socioeconomic status, and the environment. It was widely appreciated that patients' clinical care was impacted by several nonclinical factors, including health insurance coverage, transportation issues, employment, unstable housing, mental health, food insecurity, education levels, and health literacy.

One specific focus was the employment status of people with PAH: the authors recognized that work provides a sense of self-worth, keeps individuals active, and creates a routine that can help patients manage their condition. PAH can be an isolating disease, and work can offer a sense of normalcy and connection that can improve mental well-being. Moreover, working as part of a team towards a common goal can permit patients to focus on something other than illness, helping manage both physical and emotional health. However, work also offers challenges—including physical demands, rigid hours, inflexibility regarding time off for medical appointments—which can directly impact health outcomes. Jobs that require physical exertion or long hours can worsen symptoms such as shortness of breath and fatigue, limiting the ability of people with PAH to engage in physically demanding work. Simple tasks, such as walking short distances or standing for extended periods, may exhaust a patient, pushing them to seek sedentary roles that require less exertion. Often, patients cannot easily transition to less physically strenuous jobs, and the search for suitable work often adds stress to their lives. Flexibility in work hours, the ability to take time off for medical appointments, and options for remote work are all essential in helping balance employment with health needs. A supportive work environment and appropriate accommodations are invaluable to permit people with PAH to manage their condition effectively while continuing working and maintaining financial independence. Conversely, not all workplaces are willing or equipped to make these accommodations, which can intensify stress and anxiety sometimes worsening health outcomes, and also can limit employment options for people with PAH.

Implications. Experienced PH physicians across the US were highly aware of SDH and understood how inequities in clinical care were often related to patients' socioeconomic factors. Moreover, adverse SDH in patients with PAH were thought to correlate with poor health outcomes. This study could be helpful in informing a future PH practice model that includes screening for SDH (e.g. housing instability, food insecurity, transportation, financial resources strain, exposure to violence, childcare, and behavioral / mental health) using a specific SDH questionnaire, thus better integrating patient voices into their care, and improving our ability to address social and economic inequities through robust health policy initiatives.

The article contextualizes the discussion of PAH and employment in broader themes of social drivers of health, such as food and housing insecurity, transportation, substance abuse, and family support. It underscores the need to address social and economic inequities through robust health policy initiatives.

Contributed by:
Saeid Maghsoudi MSc, Graduate Student, U Manitoba, Winnipeg; Ashok Pandey BSc Candidate, U Guelph; and Sanjay Mehta MDCM, FRCPC, Founding Board Member / Past Chair / Eternal PHriend of PHA Canada, UWO, London

2024 Pediatric PH Research Scholarship Recipient

PHA Canada is proud to support the growth of the research community by awarding scholarships to outstanding trainees working in the field of pulmonary hypertension (PH). In early 2024, we introduced the PHA Canada Bell Family Pediatric PH Research Scholarship, an initiative specifically created to support pediatric PH research through the generous donation of Darren Bell. This scholarship honours the memory of Darren's son Dylan, who sadly passed away from pulmonary arterial hypertension (PAH) in 2007. Through this scholarship, we aim to advance research that will improve the lives of children living with PH.

Mahnaz Nazari

We are excited to announce that Mahnaz Nazari is the inaugural recipient of the PHA Canada Bell Family Pediatric PH Research Scholarship. Mahnaz is a third-year Ph.D. candidate in the Cellular and Molecular Medicine program at the Ottawa Hospital Research Institute, working in Dr. Bernard Thébaud's laboratory. She earned her Bachelor's in Cellular and Molecular Biology from Alzahra University, Iran, in 2016, and her Master's in Laboratory Hematology and Blood Banking from Tabriz University of Medical Sciences, Iran, in 2020.

Her current research focuses on developing treatments for extremely premature babies with pulmonary hypertension (PH). Her preliminary studies have shown promising results in using cell-based therapies in a neonatal lung injury mouse model. The goal is to create a cell-free, off-the-shelf product for clinical trials, offering new hope for premature infants with PH.

Project: Cell-Based Therapies for Pulmonary Hypertension in Premature Infants with Bronchopulmonary Dysplasia

Mahnaz Nazari's PhD project explores cell-based therapies for bronchopulmonary dysplasia (BPD) and associated PH in extremely premature infants. Using umbilical cord blood-derived endothelial colony-forming cells (ECFCs) and human induced pluripotent stem cell-derived vasculogenic progenitor cells (VPCs), along with their extracellular vesicles (EVs), she aims to provide pre-clinical data for clinical translation. Collaborations and advanced techniques like multiplexed scRNAseq will support a pre-CTA application to Health Canada, leading to a Phase I trial to assess the safety and feasibility of these therapies in neonatal PH.

Learn more about PHA Canada's
scholarship recipients and their research projects at
phacanada.ca/scholarship

Scholarship Recipient Research Update

In early 2024, PHA Canada proudly awarded its inaugural Bell Family Pediatric PH Research Scholarship to Mahnaz Nazari, a third-year PhD candidate in the Cellular and Molecular Medicine program at the Ottawa Hospital Research Institute. Working under the supervision of Dr. Bernard Thébaud, Mahnaz is leading groundbreaking research to develop cell-based therapies for premature infants suffering from pulmonary hypertension (PH), particularly those with bronchopulmonary dysplasia (BPD).

A critical contribution of cardiac myofibroblasts in RV failure and the role of UCP2 SNPs in the predisposition to RV decompensation in pulmonary arterial hypertension

The main driver of mortality and morbidity in pulmonary arterial hypertension (PAH) is the function of the right ventricle (RV) rather than the degree of rise in mean Pulmonary Artery Pressure (mPAP)[1,2]. However, the mechanism driving the transition from compensated (cRVH) to decompensated right ventricular hypertrophy (dRVH) in pulmonary hypertension (PHT) is unknown. We hypothesized that a transition from cardiac fibroblasts (cFB) to cardiac myofibroblasts (cMFB) underlies this mechanism.

In the monocrotaline-rat PHT model, we separated cRVH from dRVH based on strict catheterization and echo criteria: dRVH had decreased cardiac output, increased right atrial pressure, decreased tricuspid annular plane systolic excursion (TAPSE) and increased RV end-diastolic diameter compared to cRVH. In isolated hearts, RV systolic pressure was lower in dRVH but in isolated cardiomyocytes (CM), contractility (sarcomere shortening) was not, pointing to a non-cardiomyocyte cause. The number of cMFB was not different between cRVH and Control, but dramatically increased in dRVH. Mitochondrial respiration was lower in dRVH cMFB than cRVH cFB.

The transformation of cFB into cMFB has been shown to depend on decreased mitochondrial calcium (mCa++)[3], which decreases the activity of mitochondrial enzymes and the production of diffusible metabolites (like citrate) that regulate the epigenetic changes involved in the change of cell identity. mCa++ was decreased from normal RV to cRVH to dRVH c(M)FB, while it was not different in CM. This was due to concordant differences in the methylation of mitochondrial calcium uptake 1 (MICU1), a component of the mitochondrial calcium uniporter (MCU), as well as the expression and activity of its methyltransferase protein arginine methyltransferase 1 (PRMT1)[4]. The other component of MCU is uncoupling protein 2 (UCP2). Its expression decreased from normal RV to cRVH to dRVH c(M)FB (but not CM). To determine the role of UCP2 in the regulation of mCa++ independent of MICU1 methylation, we used cFB from wild-type, UCP2 heterozygote and KO mice, and showed a gene dose-dependent decrease in mCa++ as UCP2 levels decrease.

In a cohort of 30 patients with human RV tissues, dRVH (based on clinical criteria) had increased cMFBs compared to Control and cRVH. Cytoplasmic PRMT1 was increased and UCP2 was decreased from Control to cRVH to dRVH c(M)FB. UCP2 protein level correlated negatively with the presence of a loss-of-function SNP (rs659366) and positively with TAPSE. In a subgroup that had Echo and right heart catheterization data within 2 days, carriers of the UCP2 SNP had decreased TAPSE (i.e. RV function) compared to non-carriers with similar mean PA pressure.

Overall, Our data point to a change of cell identity (cFB to cMFB) in the RV as the basis of RV decompensation. UCP2 SNPs may be promising biomarkers for RV failure in PHT patients. However, further research and a larger cohort are needed to confirm this finding.

Contributed by: Mahnaz Nazari, Recipient of the PHA Canada Bell Family Pediatric PH Research Scholarship.

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