PULMONARY HYPERTENSION ASSOCIATION OF CANADA

L'ASSOCIATION D'HYPERTENSION PULMONAIRE DU CANADA

CONNECTIONS

The Official Magazine of the Canadian PH Community

Special Feature:

Pediatric PH

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

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For subscription inquiries and information, please contact us at 1-877-774-2226 or at connections@phacanada.ca.

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Editor in Chief
Kimberly Brunelle, Director, Programs

Contributing Editor
Robyn Kalda, Manager, Research & Policy

Contributing Proofreader
Robyn Kalda, Manager, Research & Policy

Translation
Jeremy Durand, Manager, Marketing and Communications

Art & Design
Jeremy Durand, Manager, Marketing and Communications

Printing
BP Media

Contributors
Angela Bates
Violet Cairns
Emily Cizman
Donna Clark
Elayna Clark
Pierce Colpman
Robyn Kalda
Gabriel Libuna
Brinley Marks
Georgiana McAlpine
Nicole Moores
Talitha Moores
Clare Neilson
Lara Presotto
Victoria Presotto
Andrea Gore-Ramsey
Felix Ramsey
Zoe Reeves
Janette T. Reyes
Olivia Zloty

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Celebrating the strength, spirit, and voices of young people living with pulmonary hypertension

Hello PHriends,

As spring fades into summer and the days grow longer and warmer, there's an unmistakable energy in the air. It's a season of growth, renewal, and connection—a perfect time to embrace fresh starts, new perspectives, and the resilience that continues to shine throughout our community.

We're proud to bring you a very special edition of Connections Magazine, dedicated to the youngest voices in our community—children and youth living with pulmonary hypertension (PH)—and to the caregivers, healthcare professionals, and researchers who walk beside them. Their stories remind us that even in the face of challenges, resilience, courage, and joy can bloom like wildflowers in the sun. While PH is already a complex and life-altering condition, its impact in childhood and adolescence brings unique emotional, developmental, and medical challenges. This issue is a tribute to the strength and spirit of these young people and their families, who navigate daily life with courage, creativity, and an incredible capacity to adapt.

In this issue, we are also thrilled to welcome some new (and not so new) faces to the team. We welcome Gail Nicholson as a new Board Director representing the PH nursing community. Gail is a longtime member of the PH clinic in Calgary and brings invaluable frontline experience to our Board. We're also thrilled to introduce Shelley Grogan as our new Director of Fund Development. Shelley brings over 16 years of experience in the charitable sector and hails from Newfoundland, helping us round out our national team. We're proud that the PHA Canada team is now truly coast-to-coast!

At PHA Canada, our mission to empower the Canadian pulmonary hypertension community drives everything we do. In this issue, we're proud to share how our recent work and initiatives have brought that mission to life.

Advocacy
We launched our Time Matters PAH campaign last fall and have been busy engaging with decision makers, politicians, and community leaders to advocate for the swift and successful negotiation of a price for sotatercept.

In May 2025, we marked World PH Day with powerful in-person meetings in St. John's, Newfoundland and Winnipeg, Manitoba, where patient and caregiver advocates met with elected officials to raise awareness about pulmonary arterial hypertension (PAH) and the urgent need for equitable, timely access to treatment. These discussions helped further amplify the goals of our Time Matters PAH campaign and are a testament to what happens when lived experience meets political engagement. We thank everyone who shared their stories, spoke truth to power, and helped put PAH on the policy agenda.

Awareness
We reflect on the incredible energy and impact of PH Awareness Month 2024, when patients, caregivers, and healthcare professionals nationwide came together to raise awareness and funds. Our annual 6-Minute Walk for Breath was once again a highlight, uniting participants of all ages in a virtual movement challenge to support PHA Canada's programs and services. Whether you walked, shared, or donated, your actions helped make PH more visible and gave a voice to those affected by this rare disease. Thank you for every step you took to help make a difference.

Research
Our Research Corner highlights recent developments from the 7th World Symposium on Pulmonary Hypertension, where pediatric PH was given well-deserved attention. These updates are paired with insights from Canadian PH specialists working to ensure that pediatric care reflects the unique realities of young patients and their families.

Education
As we celebrate everything we've achieved together over the past year, we also look forward to the opportunities ahead. One of the most exciting is our upcoming 2025 PH Community Conference, September 11-13 in Toronto. This gathering will bring together patients, families, healthcare providers, and researchers for three days of learning, connection, and inspiration. Whether you're attending for the first time or returning to reconnect with familiar faces, we can't wait to welcome you.

Support
Of course, none of this work would be possible without the leadership of our volunteers. From the families who contributed to this magazine, to the advocates, fundraisers, peer supporters, and Board members who guide our work year-round—thank you. Your commitment drives every program we offer and every milestone we reach.

Thank you for reading this issue, sharing your time, and being part of this incredible community. We hope the stories and insights you find in these pages bring you knowledge, comfort, and connection.

With warmth and gratitude,

Team PHA Canada

Memo:
Inside PHA Canada

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

Shelley Grogan
Director, Fund Development, Bay Roberts, NL

Shelley recently joined PHA Canada as the Director, Fund Development. With over 16 years of experience in the charitable sector, she specializes in creating sustainable fundraising strategies to support meaningful causes. Shelley has worked with both grassroots organizations and large national charities, helping to secure funding for vital programs and services. At PHA Canada, she is committed to strengthening donor engagement and building support for the PH community.

Gail Nicholson
Board Director, Calgary, AB

Gail Nicholson is a registered nurse based in Southern Alberta and a longtime member of the Canadian pulmonary hypertension (PH) community. She began her nursing career in 1988, working in medical-surgical and intensive care before joining the Southern Alberta PH program in 2007, where she has focused exclusively on pulmonary arterial hypertension (PAH) care. Gail has collaborated nationally through the Canadian PH Professionals Network and internationally on clinical papers to advance best practices in PH nursing. In 2025, she joined PHA Canada's Board of Directors, bringing her clinical insight, dedication to patient-centered care, and passion for improving the lives of those affected by PH.

Subscribe now

Looking to learn more about pulmonary hypertension? Visit PHA Canada's YouTube channel! You'll find a wide range of content, including educational webinars, expert interviews, event highlights, and inspiring stories from those living with PH.

Subscribe now at www.youtube.com/@PHACanada and stay informed, empowered, and engaged.

Marking the End of a Chapter: Thank You, Ambassadors

We thank PHA Canada's Ambassadors with heartfelt appreciation for their dedication, leadership, and lasting contributions to the pulmonary hypertension (PH) community.

Throughout the Ambassador Program, these passionate volunteers gave their time, voice, and energy to support others and raise awareness. Whether speaking at events, mentoring fellow patients, or sharing their personal experiences, Ambassadors helped foster connection, understanding, and hope across the country.

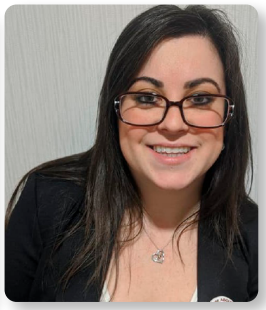
Each Ambassador brought compassion and courage to their role, offering encouragement to those newly diagnosed, advocating for better awareness, and helping others feel less alone. Their efforts helped strengthen our community and advance our shared mission.

As we close this chapter, we're excited to share that a new program—shaped by everything we learned from the Ambassador Program—is on its way. This next evolution will build on the foundation Ambassadors helped create and will offer new opportunities for community members to get involved, contribute, and lead. We can't wait to share more soon.

On behalf of PHA Canada and the entire PH community, thank you. Your contributions have truly made a difference, and we are grateful for everything you've done to support and inspire others. Stay tuned—the next chapter is just beginning.

With deep thanks,
PHA Canada & the PH Community


Lindsay Forsyth Brochu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Kathy Downey


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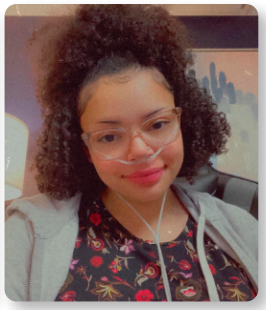
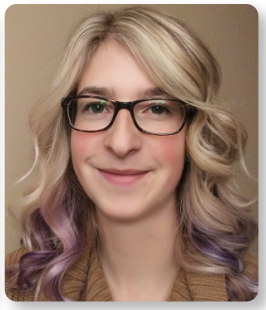

Brinley Marks


Jessica Marks-Cullum


Jane Sernoskie


Susan Silver

2025 PH Community Conference
Driving Change, Inspiring Hope

September 11-13 in Toronto - in-person or virtual

Register now at
phacanada.ca/conference

September 11-12	September 12	September 13
Medical Think Tank	Evening Cocktail Reception	Patient & Family Symposium

Join us at PHA Canada's 2025 PH Community Conference to explore how we can drive meaningful change and inspire hope across the PH community. This year's event will feature powerful keynote speakers, interactive workshops, and engaging panel discussions—all designed to empower you with new strategies, the latest research, and stories of resilience and innovation.

"This conference was really life changing for me. I have never felt so connected to people. I learned so much at all the small groups, as well as at the larger group discussions at the conference. I just felt happy from the inside out."

Stacy Physick
Living with PAH, Airdrie, AB

Your Community

This edition's Community section is a powerful reminder of how united action can create real change. Over the past year, our community has rallied together in countless ways—from gathering for the 6-Minute Walk for Breath and connecting at Pulmonary Hypertension Awareness Month events, to advocating for sotatercept on the provincial level and during World PH Day. Whether raising awareness through the Power of 6 campaign, meeting elected officials at Queen's Park, or writing letters as part of the Time Matters PAH campaign, your dedication continues to shape the future of care for people affected by PH. Every conversation, every step, and every story brings us closer to making pulmonary hypertension impossible to ignore.

PH Awareness Month:
Life in Purple

Every November, PH Awareness Month unites people living with pulmonary hypertension (PH), their loved ones, and care providers to spread awareness and push for better care. Last year, our community came together to educate the public, share personal stories, and advocate for essential treatments. Through social media, storytelling, and advocacy campaigns, we made a lasting impact. Learn more at phacanada.ca/pham.

Raising Awareness

PH is a rare and complex disease that is often misdiagnosed. To bring more visibility to the condition, we used the #LifeInPurple campaign, sharing patient experiences and the realities of living with PH. Supporters helped spread awareness through the Power of 6, telling six people about PH and inspiring them to do the same.

We also highlighted our report on the Socioeconomic Burden of PAH in Canada, shedding light on the financial and emotional challenges PAH patients face. By sharing this research, we increased understanding and support for those affected.

Advocating for Change

Our #TimeMattersPAH campaign focuses on securing public access to sotatercept (Winrevair™)—a breakthrough treatment for pulmonary arterial hypertension (PAH). With funding decisions pending, our community continues to raise its voice to demand timely access to this life-changing therapy. By engaging policymakers and rallying public support, we reinforce our mission to ensure all Canadians with PH receive the care they need.

Community Events

The highlight of PH Awareness Month 2024 was the community gatherings held in Vancouver, Winnipeg, London, and Ottawa. These events, along with PHA Canada's Annual 6-Minute Walk for Breath, brought the country together to raise awareness and celebrate the strength of Canada's PH community. Thank you to all the event organizers.

Vancouver, BC

London, ON

Ottawa, ON

The 6-Minute Walk for Breath

PHA Canada's Annual 6-Minute Walk for Breath returned on November 23, 2024, during PH Awareness Month, uniting participants from coast to coast in a powerful demonstration of community spirit. Whether walking together in local gatherings or joining virtually from home, individuals and teams showed up in force to honour those affected by pulmonary hypertension and to spread vital awareness. This year's event was a tremendous success, raising **over $55,000** and setting a new fundraising record. Thank you to everyone who participated and helped make this event so impactful—your efforts are bringing us closer to a better future for the PH community.

Top Fundraising Teams

Once again, **Team Ottawa** took the top spot, raising an incredible $12,173! Led by PH patient Jane Sernoskie, this unstoppable team continues to show unwavering dedication to the PH community. Close behind, the **Mayville Movers** rallied together to raise an impressive $12,110 in honour of Nicole and Michael Mayville's daughter, who was diagnosed with PAH in 2020. Your support is truly inspiring! Rounding out the top three, **London Purple Power**, led by PHA Canada Director Marion Roth, raised an outstanding $11,283.60—a powerful show of commitment and strength.

A heartfelt thank you to these incredible teams and to everyone who participated. Your efforts make a real difference in the fight against PH!

Top Individual Fundraisers

1. Michael Mayville - $10,005.00
2. Donna Downes - $4,379.90
3. Michael Pohanka - $4,009.20
4. Glenda J. Cracknell - $3,743.40
5. Jo-Anne Mainwood - $2,719.90

We express our heartfelt appreciation to the incredible individuals and teams who have fueled our efforts in the fight against PH.


Michael Mayville and his family


Donna Downes


Michael Pohanka

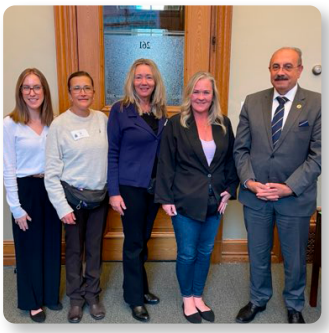

Glenda J. Cracknell


Jo-Anne Mainwood

Advocating for PAH Patients at Queen's Park

In a powerful display of advocacy, **PHA Canada advocates gathered at Queen's Park** to raise awareness of **pulmonary arterial hypertension (PAH)** and push for better access to care and life-changing treatments. Over the course of our advocacy days, we met with key Members of Provincial Parliament (MPPs) to share the urgent need for timely, funded access to sotatercept (Winrevair™) and improved support for Ontarians living with PAH.

Throughout the week, we had the honour of being recognized in the Ontario legislature by MPPs Nina Tangri, John Fraser, and France Gélinas, who acknowledged the importance of supporting the PAH community. We also had the opportunity to meet with several Members of Provincial Parliament (MPPs), including Sheref Sabawy, Andrea Hazell, France Gélinas, John Fraser, Anthony Leardi, and Monique Taylor. Their willingness to engage in discussions about PAH and the challenges faced by patients demonstrated their commitment to improving access to treatment in Canada.



These advocacy days at Queen's Park were a crucial step in amplifying the voices of those living with PAH and ensuring that policymakers understand the urgent need for action. A huge thank you to our PHA Canada advocates and to the Ontario MPPs who took the time to meet with us. Your support brings us closer to a future where every Canadian affected by PAH has access to the treatments they need to live longer, healthier lives.

Learn more about our advocacy efforts at phacanada.ca/advocate.

Patients Unite for World PH Day

This World Pulmonary Hypertension Day, our message was loud and clear: Time Matters for Canadians living with pulmonary arterial hypertension (PAH). On May 5, advocates met with decision-makers, raised awareness about PAH, and advocated for timely access to life-changing treatments like sotatercept (Winrevair™). Through heartfelt storytelling and strategic outreach, our community made PAH impossible to ignore.

St. John's, Newfoundland and Labrador

Our advocates in St. John's met with Members of the House of Assembly (MHAs) Paul Dinn, Jamie Korab, and Barry Petten, as well as Alicia Chenard, Executive Assistant to the Minister of Health and Community Services. These meetings were a powerful opportunity to share why public access to sotatercept matters—urgently. Special thanks to advocates Sonya Collins, Heather Marrie, and Denise Rumbolt, who spoke bravely about their experiences with PAH and why every moment counts. Their voices helped put a human face to our message and reinforced the need for immediate provincial action.

Winnipeg, Manitoba

In Manitoba, our advocates met with Hon. Uzoma Asagwara, Minister of Health, Seniors, and Long-Term Care, and Carla Compton, Member of the Legislative Assembly (MLA) for Tuxedo, who welcomed the group with a formal introduction in the Legislature. We also held conversations with representatives from both the Progressive Conservative and Liberal parties, ensuring cross-party engagement. Our heartfelt thanks to advocates David Christiansen, Cindy Hayman, Jo-Ann McKenzie, Christine Smilski, Tanya Stinson, Andrea Toews, Chrissy Wilder, and Rodney Wilder. Together, they painted a vivid picture of life with PAH and the hope sotatercept represents.

Time Matters PAH:
Advocate for Sotatercept in Canada

We're rallying the PAH community to urge provincial and territorial health leaders to prioritize and expedite public funding for sotatercept (Winrevair™)—a groundbreaking new therapy for pulmonary arterial hypertension (PAH).

Approved by Health Canada in August 2024, sotatercept is the first activin signaling inhibitor approved for long-term PAH treatment. It's recommended by Canada's Drug Agency (CDA) for public funding alongside existing therapies for adult patients with WHO Functional Class II or III PAH, when prescribed by a PAH specialist. After months in limbo, sotatercept is now in active price negotiations through the pan-Canadian Pharmaceutical Alliance (pCPA)—but patients are still waiting.

And PAH patients cannot afford to wait.

#TimeMattersPAH

How You Can Get Involved

1. Write to Your Health Minister: Use our template letter to urge your provincial or territorial health minister to quicken 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 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

Special Feature

Pediatric PH

In this special feature, we shine a light on the real experiences of children and youth living with pulmonary hypertension and the parents who walk beside them every step of the way. Through honest reflections, heartfelt stories, and practical insights, families share their journeys—from diagnosis to daily routines, from navigating school to preparing for the transition to adult care. These are the stories of our littlest warriors and the parents who support them with love, resilience, and determination. Each voice in this section reminds us that while a PH diagnosis brings many challenges, it also reveals remarkable strength. Together, these stories form a powerful portrait of courage, connection, and hope within the pediatric PH community.

Growing Up with PH and Facing It Together

Felix is 12 years old and has been living with pulmonary arterial hypertension (PAH) since he was 6 years old. PHA Canada spoke with Felix and his mom, Andrea, about their diagnosis journey. Felix is currently on dual oral therapy and uses oxygen at night and when traveling.

When Felix was six years old, his mom, Andrea, noticed something wasn't right. "He started taking these really deep breaths all the time, even when he was just sitting still," she remembers. At first, they thought it might be anxiety—he had just started kindergarten, and their family had recently moved from Denmark to Virginia. But Andrea had a gut feeling that something more was going on.

When they finally met with a pediatrician, Andrea explained Felix's medical history—his time in the neonatal unit as a baby, his trouble gaining weight, and frequent illnesses. Luckily, the nurse they saw had experience in cardiology. She referred them for an echocardiogram, and that's when everything changed.

"The doctor came in, sat down, took off his hat, and told me that Felix had pulmonary hypertension," Andrea recalls. "I was in shock. I had no idea what PH was, but the doctor's reaction told me it was serious. By the time I got home, I couldn't even remember the name of it—just that we finally had an answer."

Life with PH: "It Doesn't Really Slow Me Down"
Felix doesn't let PH define him. He enjoys grade 6—especially science and math—and is an active kid. "I take skating lessons, I'm on the volleyball team, and I play baseball. I usually play in the outfield, but I also like shortstop and third base," he says. Sometimes, he has to take breaks in gym class, but otherwise, PH doesn't stop him from doing what he loves. When friends ask about his condition, he keeps it simple: "I just kind of say that I have this thing where it's harder to breathe sometimes, and I need to take breaks."

Andrea admires Felix's resilience but knows that managing PH is an ongoing challenge. "Going from no medications to suddenly having to take them every day was a big adjustment," she says. "But Felix has been on oxygen for most of his life, so it's just normal for him." They travel to Toronto for specialist appointments once a year. "It's a fun trip—I don't really mind it, but I don't love it either," Felix shrugs. "But I look forward to going to Basil Box whenever we're there. The Pad Thai is really good. Or sushi!"

A Family's Journey
Felix isn't navigating PH alone—his younger brother, Max, is right there with him. "Sometimes there's tension," Andrea admits. "Felix gets to stay home from school more often, and Max doesn't think that's fair. But we try to keep things balanced." The boys are close, even sharing a room. In fact, when Felix is away at a sleepover, Max finds it hard to sleep without the familiar sound of his brother's oxygen concentrator. "He's a very supportive little brother."

For Andrea and her husband, finding support has been key. "The online PH community has been a lifesaver. It's such a complex condition that no one else understands, so having other parents to connect with is huge."

Advice for Other Families Facing PH
Looking back, Andrea has learned to take things one day at a time. "That's the best advice I ever got, and it's easier said than done. You don't know how things will go—you just have to wait and see, which is so hard. But don't be afraid to ask for help. When we were first diagnosed, we went to family counselling, and it was life-changing."

Her biggest hope for the future? "I hope new medications continue to become available in Canada. Every PH patient deserves access to the treatments they need. And more than anything, I just want Felix to keep being awesome."

If you ask Felix, he'd say he's doing just that. He's playing baseball, acing math class, or battling it out in Brawl Stars—PH may be part of his story, but it doesn't write the whole book.

Contributed by: Felix Ramsey, living with PAH since 2018, and his mom, Andrea Gore-Ramsey, Halifax, NS, as interviewed by Kimberly Brunelle at PHA Canada

Adapting to the Unexpected: A Family's Journey Through PH

Victoria, diagnosed with pulmonary arterial hypertension in 2023, is on dual therapy. Her mother, Lara, talks to PHA Canada about their family's PAH journey.

Diagnosis
When four-year-old Victoria was hospitalized for what seemed like a routine virus, her parents never expected a life-changing diagnosis: pulmonary arterial hypertension (PAH). It was definitely a shock," says Lara, Victoria's mom. "Looking back, we can spot things, like when she'd ask to be carried halfway to the park, or how much she loved to sleep, but nothing stood out."

"We were scared," Lara says, recalling how they kept Victoria indoors, away from social activities, in the early months after her diagnosis. "We didn't go to birthday parties or playdates. It was a very isolating time." Lara took Victoria out of school, and her father took a leave of absence from work to stay home with her and protect her health. "Being an only child, the isolation was hard on her. She spent a lot of time with us, and not so much with other kids," Lara recalls. "That was probably the most difficult part—keeping her away from other children."

Finding Balance and Hope
Now six, Victoria is thriving on a regimen of bosentan and sildenafil. Over time, the family carefully reintroduced social activities. Victoria has returned to school, under the watchful eye of the school nurse. "You find your new rhythm," Lara reflects. "You learn what works for your family. While things are still challenging, we've come to a place of balance."

The Make-A-Wish Foundation granted Victoria's wish to go to Disney World. "It was incredible," Lara says. "Victoria felt like a princess. It was magical. She saw other kids there—some in wheelchairs, some visibly ill. It helped her realize that she's not the only one facing something tough. That was powerful for her."

Advice for Other Parents
Lara's advice to other parents is heartfelt: "You are not alone." She encourages families to seek support through programs like PHA Canada's PH Buddy Program, which pairs newly diagnosed individuals and families with others who understand. "Even just knowing someone else out there truly gets it—that's powerful," Lara says. "Programs like the PH Buddy Program can give you a sense of belonging when you need it most."

Contributed by: Lara Presotto, mother and caregiver to Victoria, Toronto, ON, as interviewed by Kimberly Brunelle at PHA Canada

Q&A with Victoria:

Q. What's your favorite thing to do for fun?
A. Making rainbow loom bracelets, making slime, and playing with friends at school.

Q. What do you not like about going to the doctor?
A. "Echo takes too long."

Q. What's something that helps you when you have to take medicine or use oxygen?
A. "I take a deep breath and take my meds."

Q. What's something fun you would love to do one day?
A. "Go to the moon."

Contributed by: Victoria Presotto, living with PAH since 2023, Toronto, ON

Finding Her Flock: How Olivia Thrives with PH

Olivia is 14 and lives in Sundre, Alberta. She was diagnosed with PAH in 2014. She is currently on triple therapy as well as sub-q remodulin. PHA Canada recently spoke to Olivia about her experience in the 4-H club along with her adorable lamb, Atlas.

Q: Tell me about 4-H! What is it all about?

Olivia: Our 4-H club is Cowboy Trail 4-H Multi Club, and I am a member along with my two brothers. We are all raising sheep! My brothers have Twig and Cheddah, who are market lambs, and I'm raising an ewe named Atlas. We're there right when they're born, and then we raise them and train them for the show ring.

We live on a farm, so we get to bring the animals home and work with them. The 4-H club offers lots of other opportunities for kids, like sewing, cooking, archery, and photography. There's even raising a dog or a cat.

Q: So is it just about raising and training an animal?

Olivia: There's lots of community service and public speaking. We gave speeches in front of the judges and at the local seniors' center. A lot of the seniors did 4-H when they were kids, so it kind of brought them back into it.

Then there's multi-judging. We spend a full day judging—it could be something as simple as judging which cookie looks the best—but we go in front of a judge and present why we think that. The idea is that as adults, we'll be able to go into a boardroom and justify our decisions.

Q: You've won some awards—congratulations! What do judges usually look for in a winning sheep? Is it about your skills as a handler, the quality of the sheep, or a bit of both?

Olivia: For the market lamb trophies, it's how much meat is on them, their condition, and their health. How does it look? How's the build of it? Does it have good muscle strength?

Then there's showmanship. I have to walk Atlas around the ring with her halter on and compete. They even have us switch places to see if your lamb stays still and behaves in the ring with somebody else. You're proud of your little animal—to watch them grow and train them.

Q: Have they made any accommodations for you?

Olivia: They make sure I have places to sit and rest. During public speaking, I got dizzy, so they made sure I could sit down and rearranged the order so I didn't have to go first. Now I have a little collapsible stool I take everywhere. If we're walking somewhere, I might be driven. When we went sledding, they drove me up to the top of the hill over and over so I didn't have to climb it, and I didn't have to miss out.

Parents aren't allowed in the show ring, so I've had to advocate for myself and let my peers help me out. I wanted to share my story with the PH community to show kids that they can find different activities that work for them. Just because they have PH doesn't mean they can't do activities!

Q: What happens to your animals?

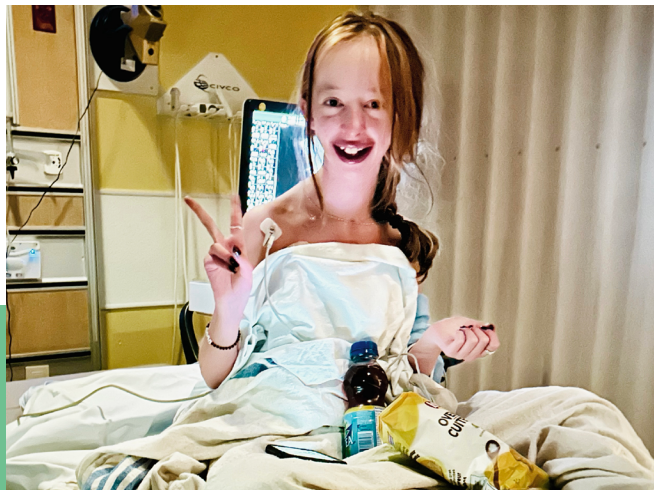
Olivia: Because Atlas is a female, I get to keep working with her. Next year, I'll breed her and show her and the babies, and the year after, I'll have my own flock. My brothers' male lambs are market lambs and will be sold.

Our club is trying to give back to our community, so we will donate a portion of our profits to a charity of our choice. My brothers are going to donate to PHA Canada. We want to sell their animals, raise awareness for PH, and raise some money for PH. At the show, the sheep are all on display in their own stalls with their stall cards and information. We thought we could put a poster on them with information about PHA Canada and maybe a QR code that people can scan to donate. Maybe having a charity might make them spend more, which means we could donate more, too, right?

Contributed by: Olivia Zloty, living with PAH since 2014, Sundre, AB, as interviewed by Kimberly Brunelle at PHA Canada

Olivia and her brothers raised over $672 for PHA Canada. Thank you!

Turning Pain into Purpose: A Family's Fight Against PH



Talitha, 15, and her mother Nicole, talk about resilience and their annual fundraiser for the PH clinic at the Stollery Children's Hospital.

When my daughter Talitha was diagnosed with pulmonary hypertension (PH) at just three years old, my world changed. Learning that your child has a progressive, life-threatening disease with no cure shatters everything you thought you knew about parenthood. The future we had imagined was suddenly replaced with uncertainty, medical terminology, and a constant undercurrent of fear.

At first, we felt incredibly isolated. We struggled to find others who understood what we were going through. Hospital visits, medical procedures, and consultations with specialists became our new reality. But through it all, Talitha (my personal hero) faced every challenge with unimaginable courage.

Children often surprise us with their resilience; Talitha has been no exception. While I was overwhelmed by medical decisions and worry, she focused on simple joys—making friends with nurses, collecting hospital bracelets, or looking forward to a treat after appointments. She has tackled every hurdle with unwavering strength, showing true bravery.

Talitha's journey inspired me to transform our pain into purpose. We would use this disease to create something meaningful: Talitha's Hope 4 A Cure. What began as a small annual fundraising gathering has blossomed into a beloved community event. Each year, I am overwhelmed by the support from friends, family, healthcare workers, and even strangers. We've raised nearly $1.5 million for the PH Clinic at the Stollery Children's Hospital to help sustain critical research initiatives that might one day lead to a cure. These funds have also enabled the clinic to hire medical students, expand its team, and enhance the care it provides. Knowing that Talitha's Hope 4 A Cure continues to make a tangible difference in the lives of PH patients and their families brings profound meaning to our journey.

We've been honoured with a philanthropist award, a hospital wall naming, and a Queen's Jubilee Medal, but the real reward is bonding with other people who genuinely understand what it means to live with PH. The amazing families we've met know exactly what it's like when treatment protocols change, when symptoms unexpectedly flare, or when we experience rare moments of normalcy. It's a gift that makes this difficult journey less lonely.

To parents facing a new PH diagnosis for their child: you may feel alone, but you are not. Once you connect with other families, the burden becomes a little easier to bear. These connections won't change your child's diagnosis but will change how you experience it.

Though pulmonary hypertension entered our lives uninvited, the community, purpose, and hope it has helped us discover light our path forward—not despite Talitha's condition, but because she has taught us to face it.

Talitha's Words

Having pulmonary hypertension totally sucks. There are days when I just want to be a normal teenager without all the medications and appointments.

Even through all the hard stuff, I've found many things to be grateful for. I'm happy to have met other kids who are just like me. My PH team isn't just a group of doctors and nurses, they are like family. The medications, as annoying as they are, keep me stable enough to have good days. I am most grateful for our yearly fundraiser Talitha's Hope 4 A Cure. Being surrounded year after year by people who have joined my fight to cure PH reminds me I'm not alone.

Contributed by: Nicole Moores and her daughter, Talitha Moores, living with PAH since 2013, Fort McMurray, AB

PH Community Quotes

When you head to the hospital for appointments or tests, what are the must-have items in your hospital bag? Do you have a favorite toy, book, or snack that makes the wait easier?

I make SURE I have my phone so I can watch my favourite comfort shows like Spirit or Dolphin Tale. I take my headphones too and mom brings a charge cord. I put on my comfiest pajamas and take my 'blankie'. I also take a water bottle but the hospital has the best ice water that I like better. And I tell my friend Stephie, who works in child life, that I'm going to the hospital so she can visit me.

Zoe Reeves, 11 years old, living with CTEPH since 2018, Beaver County, AB

When I go to the hospital for a full day appointment, I pack snacks, water, lip balm, hoodie, air pods and my phone.

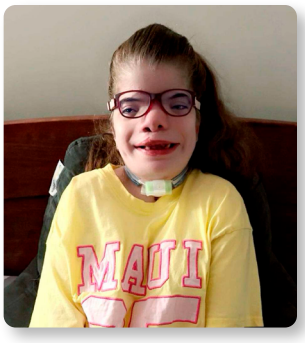
Clare Neilson, 14 years old, living with IPAH since 2012, Toronto, ON

Definitely my cell phone and phone charger, as well as my pencil case and paper to write or draw on helps to distract me. I used to bring a stuffy with me. That helped. My mom also used to make me a deal that, if I had to get blood work done, she would take me to the gift shop and buy me a new stuffy or a treat. Now, after my appointment, she always takes me to Starbucks. That's one of my favourite treats.

Elayna Clark, 13 years old, living with PAH since 2019, Cloverdale, BC

I like to bring my iPad, snacks, water bottle and a special helper stuffed animal—Marshall from Paw Patrol.

Emily Cizman, 12 years old, living with PAH since 2022, Kitchener, ON



When we head to the hospital for an appointment, we're prepared with a hospital bag stocked with essentials, such as an emergency kit for his Remodulin sub-q, wipes, a diaper, extra clothes, and some comfort items like his favorite book or toys to make the experience more enjoyable, especially during longer appointments.

Gabriel Libuna, 8 years old, living with PAH since 2017, and his mom Grace Libuna, Vancouver, BC

When I have hospital appointments, I take my Nintendo Switch to pass the time and I get excited for hot chocolate and pizza in the hospital cafeteria.

Georgiana McAlpine, 11 years old, living with PAH since 2020, Fruitvale, BC

When I go to my appointments at Sick Kids, I like going to Marnie's Lounge to play pool and make slime. I like to bring my lamb stuffed animal with me too, her name is Winnie. I also love eating the cake pops from Starbucks when my appointments are done! I won't let PH stop me from doing sports like baseball, soccer, basketball. I love to be active.

Violet Cairns, 10 years old, living with PAH since 2024, Manitowaning, ON

Starting School with Pulmonary Hypertension: One Mom's Journey

Donna Clark is an Ontario high school teacher and mom to six-year-old Xavier, who lives with pulmonary hypertension.

When your child lives with a complex medical condition like pulmonary hypertension (PH), sending them off to school for the first time feels less like a milestone and more like a leap into the unknown. That leap was overwhelming and terrifying for me, but ultimately, thanks to a supportive school and some fantastic resources, it was incredibly rewarding.

Xavier is our foster child. He's been with us since he was four weeks old, and he's now a bright, resilient six-year-old with the happiest little personality. In addition to having PH, Xavier has a global developmental delay and a genetic condition. There are a lot of layers to Xavier, which means there was a lot to consider when it came time for him to start school.

The first big hurdle was ensuring the school understood what PH is and what Xavier might need throughout the day. I was so grateful that someone from the pediatric PH community posted a link to PHA Canada's school resource guide for parents on Facebook. That resource was a game changer. It allowed me to clearly outline everything: what PH is, what symptoms to watch for, how weather impacts Xavier, and what kinds of supports he might need during the school day. Before school started, I filled in the forms, added a letter from his doctor, and met face-to-face with the principal, teacher, and ECE. We could have just emailed, but it made me feel better to meet the people who would be with my child for most of the day.

They were terrific. From day one, the staff showed compassion and a willingness to adapt. Because Xavier was still taking daily naps when he entered Junior Kindergarten, we included a note in the medical letter requesting a designated rest space. The school arranged for a cot and gave him a quiet place to nap during lunch while being supervised by an educational assistant. It was the perfect solution—not disruptive, but supportive.

Xavier's energy levels are always a consideration. His teachers have learned to read his cues. They know he might just need a break or a nap if he's grumpy or visibly tired. That flexibility has made a huge difference.

Weather accommodations have also been necessary. On days when it's extremely hot, cold, or the air quality is poor, Xavier stays indoors with an adult. At first, he struggled with that. Like any child, he didn't want to miss recess and didn't understand why he had to stay inside. But the teachers made those moments fun and routine. They never made him feel different. And over time, it just became part of his day.

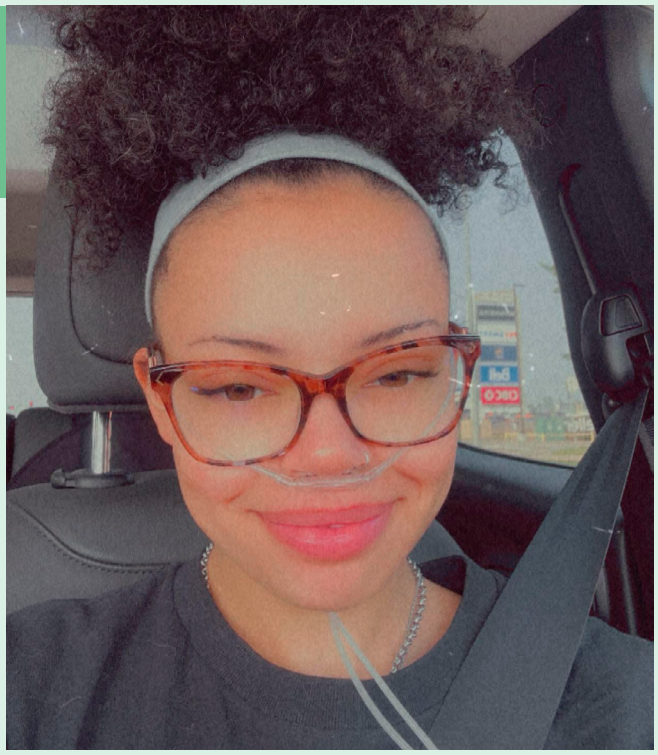
For any parent preparing to send a child with PH to school, my biggest advice is this: never be afraid to overcommunicate. You are not being a nuisance when you check in regularly or send detailed notes. You are advocating for your child. And never feel like you're giving too much information—teachers need to know who your child is so they can support them fully.

We're lucky. Xavier's school has embraced his needs with open arms. They've accommodated him not just because they have to but because they care. As an educator myself, I can tell you that makes all the difference.

Contributed by: Donna Clark, mother and caregiver to Xavier, living with PAH since 2021, Parkhill, ON

Finding My Voice:
Transitioning from Pediatric to Adult PH Care

Brinley is 20 and was diagnosed with PAH in 2020. Here, she talks about her experience transitioning from pediatric to adult care.



Imagine being 14 years old, starting your first year of high school, and suddenly being diagnosed with a terminal illness. High school is already overwhelming—going from being a big fish in a small pond to being a minnow in a lake full of bigger fish. That was me.

My high school journey started with a collapse in gym class, leading to a diagnosis of severe pulmonary arterial hypertension (PAH). It was a terrifying time, but I soon started feeling more confident. As the oldest patient in the pediatric ward, I quickly got to know all the nurses by name and found comfort in the familiar faces of my medical team. My pediatric team was kind and supportive and always made sure I had everything I needed to stay as healthy as possible. But clinic days were frustrating—my doctors discussed treatment plans and medications with my mom, without including me in the conversation. I often felt like a bystander. I wanted to be heard and have a say in my care.

Just as I had settled into my pediatric care routine, everything changed. Turning 18 meant transitioning to the intimidating adult care system, and it felt like the rug had been pulled out from under me. In pediatrics, I had a routine that gave me a sense of control—something I desperately needed after losing control over my own body at 14. Now, I had to start over.

Meeting my adult PH specialist, my mind raced. Would my doctor talk to me directly? Would I have more tests, more medications, more hospital stays? Would I have a say in my care? Walking into that appointment, I was overwhelmed and hesitant, but my fears quickly faded. My new specialist treated me like a mature young adult, addressing me directly and involving me in my care decisions. The clinic wasn't fully renovated, which oddly comforted me—maybe because, as a teenager with a messy bedroom, I was used to finding order in chaos.

My doctor handed me a consent form. It was a simple act, but it meant everything: I felt I had control over my health again. My voice mattered. I left the clinic feeling lighter, like I could finally take a deep breath. I learned I could participate in clinical trials—I'm passionate about contributing to medical advancements. I also found out that adult cardiac catheterizations are done through the arm instead of the groin, which made me nervous, but I trusted my new team to guide me through it.

The transition was nerve-wracking, but ultimately, it was empowering. I went from feeling passive in my healthcare to retaking charge of my life. Now, I have clinic visits every three months instead of every month, and my echocardiograms are far less frequent. I've even taken part in a few clinical trials.

I have PH, but PH doesn't have me. Every day, I take back a little more control, which is everything to me.

Contributed by: Brinley Marks, living with PAH since 2020, Edmonton, AB

Tips from a Pediatric Pulmonary Hypertension Nurse Practitioner

PHA Canada sat down with Janette T. Reyes, a Nurse Practitioner-Paediatrics in the Pulmonary Hypertension Service at The Labatt Family Heart Centre, the Hospital for Sick Children.

Q. Are there ways to make medication routines easier for families?

Janette. Setting phone alarms can be a helpful reminder. Using pill organizers with clearly marked a.m. and p.m. slots can make it easier to track doses. Placing medications or pill organizers visibly at routine locations such as by the bedside for morning and/or evening doses, or on the kitchen table for breakfast and dinner, can also support consistency.

Q. Are there resources you recommend to parents for emotional or practical support?

Janette. Peer support from another parent or a patient or a similar age who has a comparable PH diagnosis, treatment, and personality can be incredibly valuable for both patients and families. When appropriate, patients are referred to a pediatric psychologist or psychiatrist to support their emotional well-being and help them cope with the challenges of living with PH.

Sometimes, parents choose to bring siblings to hospital clinic appointments so they can see firsthand what their brother or sister is experiencing, which can foster empathy and understanding.

Q. Are there any school accommodations you recommend that people might not immediately think about including in their request to the school?

Janette. Children with pulmonary hypertension are more vulnerable to shortness of breath and fatigue in harsh weather conditions, so accommodations for temperature extremes are important. During very cold weather, they should remain indoors. In very hot weather, air conditioning or a fan in the classroom can help them stay comfortable.

We can arrange virtual meetings with parents, teachers, principals, and educational aides to develop a personalized plan of care. If the child is on continuous intravenous or subcutaneous therapy, I arrange a day that I can visit the school in person to speak with teachers and classmates, helping them understand the pumps and required safety that is involved. Additionally, we work closely with a Remodulin Nurse who can also provide education about the pump and safety features.

Q. Are there small things kids and parents can do to prepare for the transition to adult care?

Janette. Begin transition education around age 14 by reviewing what pulmonary hypertension is and how it leads to their symptoms. Encourage them to start learning the purpose of each of their medications. Start with simple explanations and build on that knowledge as they mature. Over time, this helps older teens become familiar with the names and doses of their medications, preparing them to take a more active role in treatment decisions and eventually managing aspects of their own care.

Q. What's one practical piece of advice you often find yourself giving to families when they first receive a PH diagnosis?

Janette. Avoid using general online searches like Google when looking for information about pulmonary hypertension (PH). Children with PH are unique, and their care needs are very much individualized. Instead, rely on trusted sources such as PHA Canada or the Pulmonary Hypertension Association, which offers accurate, up-to-date information from PH experts. A PH diagnosis can feel overwhelming, but you are not alone. We are here to support you every step of the way, taking things one step at a time and providing close, ongoing follow-up to ensure the best possible care for your child.

Contributed by: Janette T. Reyes, MN, NP-Paediatrics, Toronto, ON

Research Corner

This section features an update by Dr. Angela Bates summarizing the exciting research from the 7th World Symposium on Pulmonary Hypertension on pediatric pulmonary hypertension. We also summarize a research paper which explored whether children's bodies handle selexipag in the same way as adult bodies do, and hear from past Mohammed Family PH Research Scholarship recipient Pierce Colpman about his research looking at mitochondrial fission and pulmonary arterial hypertension.

Pediatric Updates from the 2024 World Symposium on Pulmonary Hypertension

The 7th World Symposium on Pulmonary Hypertension (WSPH) was held in Barcelona, Spain this past year at the end of June. The pediatric task force met and summarized new data and experience within our neonatal and pediatric pulmonary hypertension (PH) population that has evolved since the last WSPH in 2018. Some of the exciting new insights and recent consensus regarding the diagnosis and treatment of children living with PH around the world are being shared within the global community. This includes definitions and characterisation of PH in different pediatric age groups, current opportunities and challenges regarding risk stratification in children (which mirrors adult experience but speaks specifically to the pediatric pulmonary hypertension group), and the proposal of a new treatment algorithm which encompasses the use of new medications and non-pharmacotherapeutic therapies for pediatric pulmonary arterial hypertension (PAH) in all risk groups.

Importantly, the task force highlighted gaps in knowledge where future research efforts can be focused to continue moving the field forward, providing new tools and therapies for clinicians caring for these patients.

Cardiac catheterization

Over the past several years, there has been a shift in the definition of pulmonary arterial hypertension, as many families and patients in the pulmonary hypertension community are aware. This allows for earlier detection and intervention. As cardiac catheterization ("cath") continues to be our gold standard to diagnosis and follow PAH, there is often anxiety and risk that comes with each cath. As our knowledge of right ventricular strain (how hard the right ventricle works) evolves, alongside the use of cardiac MRI (CMRI) and cardiopulmonary exercise testing (CPET), there is a trend to do "caths" less frequently, although they remain an integral part of pediatric PH diagnosis and treatment algorithms.

Risk assessment

Risk assessment tools and scores are continually evolving in the pediatric pulmonary hypertension population and serve several purposes. The tools currently incorporate individual values from non-invasive tests, hemodynamic numbers from cardiac cath and values obtained from CMRI images.

The risk assessments help in the following ways:

1. to characterize the patient's risk for adverse outcome at any point in time and with any procedures/surgeries
2. to help clinicians monitor disease progression and the child's response to treatment, and
3. to guide treatment decisions.

Currently, World Health Organization functional class (WHO-FC), echo obtained tricuspid annular plane systolic excursion (TAPSE) and labwork value N-terminal pro-brain natriuretic peptide (NT-proBNP) are most frequently shown to correlate with outcomes in pediatric pulmonary arterial hypertension; all three of these variables have been validated in different and independent cohorts of pediatric PH patients. These variables are useful to clinicians and

As we enter an exciting new therapeutic time in pediatric pulmonary hypertension, continue to ask questions, advocate for yourselves and your families, and remember your clinical team is here to support you all along your journey.

families in predicting outcome both at diagnosis and at follow-up; as these values change over time, this information is also predictive of outcome. The hemodynamics from cath (PVRI, SVI, PACI and RAP) are helpful to predict those at 5 years who are at risk of Potts shunt, lung transplant and death, which still supports the need for cath at specific time points or if patients are not doing well clinically. CMRI right ventricular function and muscle mass also coincides with outcomes.

All these markers are clinically useful and can also be helpful in research studies as different drug trials are developed in children. How additive effects of the different markers' values affect a child's outcome continues to be studied. Keeping a low risk profile continues to be associated with better survival!

Treatment

Treatment, as many families are aware, is always evolving. Unfortunately, for children in Canada, none of the treatments are Health Canada approved. Sildenafil and bosentan are FDA approved for pulmonary arterial hypertension in children. Provincial formularies vary in terms of which drugs are on formulary and which require additional funding. Advocacy efforts continue to be part of pediatric pulmonary hypertension care!

Worldwide, treatment of pediatric PH continues to be hindered by the lack of randomized controlled clinical trials (RCTs), and it is unlikely that we will have any new RCTs for existing therapies in the near future. The challenges of drug approval include but are not exclusive to the design of pediatric clinical trials with age-appropriate reliable reproducible endpoints and adequate enrolment numbers so that results can be meaningful.

There is a newly updated proposed treatment algorithm that many of the North American and European care professionals follow. This algorithm uses a high risk/low risk stratification, applies the benefit of those patients who respond well to oxygen and inhaled nitric oxide in the cath lab, is based on a combination of real-world data as well as expert experience and extrapolation from adult clinical trials. The benefit is the inclusion of upfront combination therapy for pediatrics! We have more oral options available and hope to keep children on these therapies for as long as possible.

The task force also identified two important groups that deserve increasing attention over the next five years. First are those children with congenital heart disease that can be operated on and evaluation of surgical timing and other modifiable risk factors for PH. Second is characterizing neonatal pulmonary hypertension and understanding its differences and how we can study this population more effectively, especially in an era of increasing survival of premature babies, or those with other lung diseases.

In future

What's still missing? How do we prevent pulmonary arterial hypertension? How do we identify pulmonary hypertension even earlier with application of genetics? How do we take different groups of pulmonary arterial hypertension patients that respond differently and modify treatment algorithms, balancing quality of life and long-term outcomes? And for those already living with the disease, how does the path from diagnosis, throughout childhood and adolescence look before embarking on the next journey: living with a lung transplant?

For those parents whose children are at any point in the therapeutic pediatric PH pathway, exciting new research of drug therapy in pediatrics will be published over the next few years. Both TOMORROW (macitentan in pediatric PAH) and SALTO (selexipag in pediatric PAH) studies have completed enrollment, and we are awaiting results! The first pediatric pulmonary hypertension drug randomized control trial looking at single vs dual upfront therapy (sildenafil with or without bosentan) has also been underway.

What's on most people's minds is a new therapeutic option targeting a new pathway that has promising results in our adult friends. Sotatercept is a new fusion protein that traps ligands and rebalances pulmonary vascular balance of cell growth and death, with a goal of stopping abnormal cell proliferation, and alleviated inflammation in the vessel walls, leading to remodeling and restoration of vessel patency; adult results are very encouraging!

In pediatrics, it is currently in phase II trials (looking at dosing and side effects) but there is a plethora of real-world experience evolving in our neighbors to the south. Health Canada authorized sotatercept with a notice of compliance in August 2024.

January 31, 2025, Health Canada approved it for sale in Canada. Currently, efforts on access through Merck or through our provincial formularies are underway. Pediatrics has its work cut out for us, but we believe in our government representatives to help us safely introduce the use of sotatercept use into our pediatric drug repertoire as soon as safely possible. Stay tuned!

As we enter an exciting new therapeutic time in pediatric pulmonary hypertension, continue to ask questions, advocate for yourselves and your families, and remember your clinical team is here to support you all along your journey!

Contributed by: Angela Bates, MD, FRCPC, Pediatric Critical Care Intensivist, Stollery Children's Hospital, Edmonton, AB

As we enter an exciting new therapeutic time in pediatric pulmonary hypertension, continue to ask questions and advocate for yourselves and your families.

Research Update

Do Children's Bodies Handle Selexipag Like Adults Do?

Background. Adults with pulmonary arterial hypertension (PAH) have several approved medications that target the prostacyclin pathway, but none of them are approved for use in pediatric patients. Selexipag is one of those drugs: it's been shown to slow the progression and reduce complications of PAH in adults. For adults, we have good data on how selexipag works in the body, how effective it is, and how safe it is. This study tested the expectation that children's bodies handle the medication selexipag similarly to how adults' bodies do.

The study used a model (developed in the GRIPHON study) of how selexipag moves into, through, and out of adults' bodies, to predict appropriate doses for children. They focused on 3 weight ranges: 9 to 25 kg, 25 to 50 kg, and over 50kg, and calculated potential starting and maximum doses of selexipag for children in each weight group.

Findings. Selexipag doses were calculated to try to obtain blood levels in children similar to the range of blood levels of selexipag observed in small to normal-sized adults. The calculated selexipag doses were then tested in a clinical study of 63 children with PAH who were between 2 and 17 years old and who weighed between 9.9 and 93.5 kg. They were given increasing doses of selexipag over 12 weeks, with different starting and maximum doses based on their weight.

Researchers took blood samples from the children to measure blood levels of selexipag and related substances (known as metabolites) that are created in the body after the breakdown of selexipag. Blood levels of selexipag and its metabolites were measured one week after starting selexipag treatment and one week after reaching the maximum dose after 12 weeks.

Overall, blood levels of selexipag and its metabolites in children in all three weight groups were very close to the levels predicted by the model. Moreover, blood levels were largely the same if the children took their selexipag with water, with soft food, or dissolved in apple or orange juice.

Implications. This study confirmed that selexipag doses in children of different weights were accurately predicted by a model based on dosing in adult patients. This suggests that selexipag is handled in the children's bodies in the same way as in adults' bodies. Based on the findings of this study, the authors suggest children 2 years of age and older would be appropriate for future clinical pediatric studies of selexipag. Selexipag may become a future treatment option in children with pulmonary arterial hypertension, but the next step would be to conduct another, larger study to evaluate whether selexipag is safe and effective in children with PAH.

Contributed by: Robyn Kalda, Manager, Research & Policy

Reference: Lene Nygaard Aselsen, Anne Kümmel, Juan Jose Perez Ruixo, and Alberto Russu. Population pharmacokinetics of selexipag for dose selection and confirmation in pediatric patients with pulmonary arterial hypertension. Clinical Pharmacology & Therapeutics: Pharmacometrics & Systems Pharmacology 13(12): 2185-2195.

Scholarship Recipient Research Update

In 2021, PHA Canada awarded the Mohammed Family PH Research Scholarship to Pierce Colpman, a candidate in the Translational Medicine master's program at Queen's University. Here he describes some of his later work, which provides new insight into the development of pulmonary arterial hypertension. Colpman has also been working with Aarogya Technologies to increase accessibility to chronic disease management tools, including for conditions like PH.

Disrupted Mitochondrial Splitting May Drive Lung Vessel Overgrowth in PAH

Pulmonary arterial hypertension (PAH) is a progressive and life-threatening condition where the small arteries in the lungs become narrowed and thickened. This places intense pressure on the right side of the heart, often leading to heart failure. Scientists have long known that abnormal behavior in the cells lining these arteries, especially smooth muscle cells, plays a central role in the disease. These cells grow too much and live too long, contributing to vessel blockages and high pulmonary pressure.

In our recent study, conducted with colleagues in the Archer Lab at Queen's University, we explored a possible upstream driver of this abnormal cell behavior: mitochondrial fission. Mitochondria are the powerhouses of our cells, and they continuously split and fuse to meet the cell's energy demands and respond to stress. When this dynamic balance is disrupted, it can set off disease processes.

We focused on a protein called **MFF** (Mitochondrial Fission Factor), which helps trigger mitochondrial division. Importantly, researchers have proposed that there are at least two distinct types of mitochondrial fission:

- **Peripheral fission**, which may occur to isolate and remove damaged sections of mitochondria—a maintenance process often linked to cell stress responses.
- **Midzone fission**, where the mitochondrion divides near its center, a pattern thought to promote cell growth and proliferation.

In lung tissue from human PAH patients and animal models of the disease, we found that MFF levels were significantly increased, along with a rise in midzone fission events. This suggests that mitochondrial behavior may be skewed toward promoting the unchecked cell growth that drives the progression of pulmonary arterial hypertension.

When we blocked MFF, either through genetic techniques or a small-molecule inhibitor, the cells became less proliferative and more prone to appropriate cell death. Their mitochondria also returned to a healthier shape and function. In rats with experimental pulmonary arterial hypertension, targeting MFF improved lung structure and heart performance.

These findings suggest that mitochondrial fission, especially midzone fission driven by MFF, may contribute directly to the damaging changes in lung vessels that define pulmonary arterial hypertension. By targeting this process, future therapies might address the root causes of the disease, not just manage its symptoms.

Contributed by: Pierce Colpman, past Mohammed Family PH Research Scholarship recipient (2021)

Reference: Colpman, P., Chen, K.-H., Mewburn, J., Goodall-Halliwell, I., Das Gupta, A., & Archer, S. L. (2023). Abstract 15168: The Role of Mitochondrial Fission Factor (MFF) and Increased Midzone Mitochondrial Fission in Pulmonary Arterial Hypertension: Implications for Proliferation, Mitophagy, and Apoptosis. Circulation, 148(Suppl_1), A15168–A15168.

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