



PULMONARY HYPERTENSION
ASSOCIATION OF CANADA

2008 - 2018

L'ASSOCIATION D'HYPERTENSION
PULMONAIRE DU CANADA

A UNITED PH COMMUNITY

CONNECTIONS

The Official Magazine of the Canadian PH Community

Fall 2018 | Vol. 9, No. 2



**Special Issue:
Celebrating 10 years of
PHA Canada (Part 2)!**



**PULMONARY HYPERTENSION
ASSOCIATION OF CANADA**

2008 - 2018 L'ASSOCIATION D'HYPERTENSION
 PULMONAIRE DU CANADA
 A UNITED PH COMMUNITY

Connections is published twice a year by the Pulmonary Hypertension Association of Canada.

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

All rights reserved
 © 2018 Pulmonary Hypertension Association of Canada

Editor-in-Chief
 Michaël Robach
 PHA Canada
 Communications and
 Engagement Coordinator

Contributing Editor
 Jamie Myrah
 PHA Canada
 Executive Director

Translation
 Denise Bérubé
 Jean Dussault
 Josée Dussault
 Isabelle Lamarre
 Michaël Robach

Art & Design
 Michaël Robach

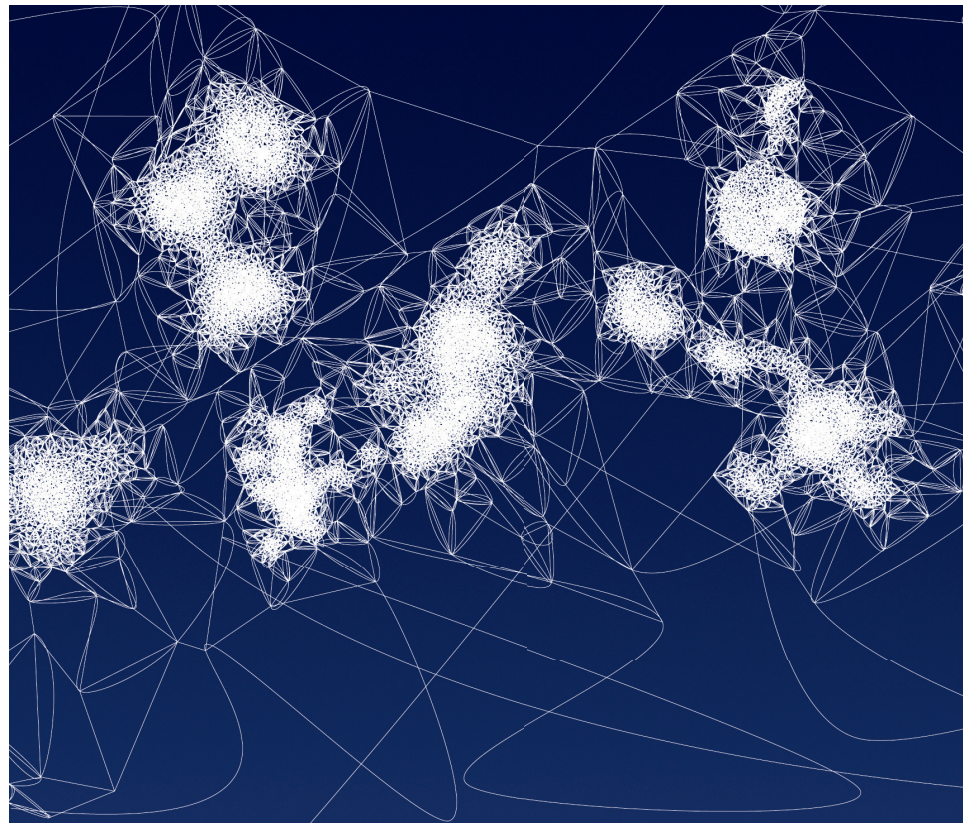
Printing
 BP Media

Contributors
 Jennifer Adams
 Jodi Ashini
 Susan Bailey
 Alice Bourgeois
 Bonnie Blenkinsop
 Dolorès Carrier
 Dr. Marc de Perrot
 Laura Donahoe
 Pat Fleming
 Olga Leal
 Lisa Lee
 Lena Legkaia
 Jo-Anne Mainwood
 Jolene Masiondz
 Dr. Sanjay Mehta
 Jamie Myrah
 Jesse Nicole
 Philip Paroian
 Sharon Proudfoot
 Dr. Steeve Provencher
 Michaël Robach
 Beth Slaunwhite
 Dr. Duncan Stewart



Cover photos (left to right, top to bottom): Stacey Jeong, Julian Carmona, Janette T. Reyes, Melanie Monds, Sabina Spataro, Adam Blanchette (& daughters), Lisa Lee, Lena Legkaia, Bal Dhillon, Dawn Dutrizac-Larose, Paul Larose, Sharon DiSanto, Dennis Guiotto, Lucas Guiotto, Michaël Robach

Unless otherwise noted, all of the photographs in this issue were contributed by authors or by PHA Canada.



ACTELION

ACTELION WOULD
 LIKE TO CONGRATULATE
 PHA CANADA ON THEIR
 10TH ANNIVERSARY.

**We celebrate your commitment to
 the PH community.**



In this Issue:

Inside PHA Canada

- 4** **Message from the Chair:**
Contributing to PHA Canada—Bigger Than Any Single Individual
by Dr. Sanjay Mehta
- 5** **Message from the Executive Director:**
Our Success Depends on You
by Jamie Myrah
- 6** **MEMO:**
Inside PHA Canada At-a-Glance

Your Community

- 8** **Your Community In Action**
by Michaël Robach
- 10** **Fundraising 101: Putting on the 2018 Ottawa**
‘Unmasking PH’ Masquerade Ball
by Susan Bailey
- 11** **Foundation HTAPQ News**
by Dolorès Carrier

Patient Stories

- 12** **My Experience Participating in a Human Stell Cell Trial**
by Jo-Anne Mainwood
- 14** **Nothing About Us, Without Us**
by Beth Slaunwhite
- 15** **Adapting to My New Normal**
by Olga Leal
- 16** **What Does PHA Canada Mean to You?**
Community Quotes

10th Anniversary Feature

- 18** **Opening Letter: Celebrating 10 Years!**
by Sharon Proudfoot
- 20** **Celebrating from Coast to Coast to Coast**
by Michaël Robach
- 22** **The Future of PH Therapies:**
Zooming In on Canadian PH Research
by Various Contributors
- 24** **PH Advocacy Through The Years**
by Jamie Myrah
- 26** **Closing Letter:**
A Physician’s Perspective on the Future of PH
by Dr. Sanjay Mehta

Medical Minute

- 28** **A National Registry for Canadian PH Patients**
Interview with Lisa Lee (NP) and Lena Legkaia

Research Corner

- 30** **FOXM1: A Potential New Therapeutic Target for**
Pulmonary Hypertension
by Alice Bourgeois

Message from the Chair:

Contributing to PHA Canada—Bigger Than Any Single Individual



This year marks a number of important milestones for the Canadian pulmonary hypertension (PH) community, as well as for me personally.

On the personal side, I completed my Specialist Fellowship in Respiriology and my training in the care of PH patients at McGill University in Montreal in 1993. As such, 2018 marks 25 years of looking after and supporting

PH patients and their caregivers, educating health care providers locally and around the world about PH, and pursuing research into the better diagnosis and treatment of PH.

On behalf of the Canadian PH community, all year PHA Canada has been proudly and enthusiastically celebrating the 10th anniversary of our founding in 2008. Through many events and celebrations, we have shared the valuable history of this incredible journey: from the initial seed of an idea around 2006, to the full realization of the dream of a national charitable organization dedicated to the vision of a better life for all Canadians affected by PH. Many individuals have been instrumental in the growth and development of PHA Canada over the past 10 years. This includes a small but incredibly passionate and dedicated team of staff, as well as countless volunteers—including past and present Board members—who have freely contributed vast amounts of their time, energy, and often direct financial support to PHA Canada.

I have had the immense pleasure of being involved in this exciting journey since the initial conception and founding of PHA Canada, and of working closely alongside many special people over the past 12 years. It was exactly because I believed—as did many others—in the importance of establishing a solid and sustainable national organization to serve PH patients and their caregivers that I became a Founding Member of the Board of Directors in 2008. Subsequently, in 2013, I agreed to serve as Chair of the Board of Directors, making a commitment in my mind of likely five years in order to continue the vital work of helping to develop and promote PHA Canada.

Today, PHA Canada has reached the exact status I hoped and worked for: it has become the de facto national voice of the Canadian PH community. PHA Canada effectively supports and educates PH patients and their caregivers across Canada, advocates on their behalf to receive optimal PH care (including publicly funded access to all 10 Health Canada-approved PH-targeted treatments), promotes medical and general community awareness of PH, and provides direct financial support to PH researchers. As such, I have decided that 2018 will be my final year as Chair of the Board of Directors.

Any transition in life can be a difficult one. Change usually involves losing or giving up something, sometimes before we may be fully ready to move on. However, there is also the intriguing promise of new and very different opportunities. Although I am stepping down as Chair, I have every intention of remaining a part of PHA Canada and will remain on the Board

as one of the Founding Board Members. Moreover, I will continue to serve on PHA Canada's Medical Advisory Committee (MAC) alongside many excellent, highly dedicated, and caring PH expert physician colleagues from across Canada. Looking around me, I am very impressed and heartened by the wealth of smart and highly skilled people who make PHA Canada hum today! I have every confidence that PHA Canada will not only remain sustainable and thrive, but will continue to grow well into the future.

Most importantly, I will absolutely continue to look after PH patients at the Southwest Ontario PH Clinic in London, Ontario. Among all of my other

responsibilities, there is nothing that is as important and personally rewarding to me as helping each individual patient and their caregivers understand, accept, and live better with their illness.

Sanjay Mehta, MD, FRCPC, FCCP
Director, Southwest Ontario PH Clinic
LHSC—Victoria Hospital, London, ON
Chair, Board of Directors, PHA Canada

“The greatest rewards come when you give of yourself. It's about bettering the lives of others, being part of something bigger than yourself, and making a positive difference.”

– Nick Vujicic

Message from the Executive Director:

Our Success Depends on You



As we have celebrated our 10th anniversary this year, PHA Canada has taken the opportunity to reflect on the purpose and people that have brought us this far. Whether you attended an anniversary party somewhere across the country (page 20), bought a special commemorative mug or tote bag (www.phacanada10.com), or contributed to this year's two special

anniversary issues of *Connections*, thank you for joining us in celebrating Canada's United PH Community!

One of this year's most noteworthy celebrations was when we honoured 11 of PHA Canada's most significant leaders of the past decade at this year's Western Regional PH Symposium in September. We look forward to sharing more with you soon from this very special occasion. It is because of the many people who have stepped up over the years to build our organization and community that we are the unified force we are today. We are so fortunate for the commitment and generosity that our volunteers, advocates, fundraisers, donors, and sponsors continue to demonstrate. It is your support that allows us to provide programs ranging from support and educational events, to advocacy and awareness campaigns, to scholarships for young researchers. It is you that keeps us moving forward.

In the last issue of *Connections*, we learned a lot about PHA Canada's history. In this issue, I invite you to gaze towards the future. To begin with, check out a new segment of the magazine on the next page for an update on some of the exciting changes occurring at PHA Canada, including an introduction to our new Ambassadors and Board Directors. These important leaders help ensure PHA Canada is working to serve you in the best ways possible, while also doing vital work to help advance our mission. Likewise, the fundraisers and PHighters featured in the *Your Community* section (page 8) show us how a little teamwork and a lot of passion for the cause can go a long way. And in *Your Stories* (page 12), three community members teach us about the power of persistence and hope. From confronting challenges of diagnosis and living with an invisible illness to contributing to advances in PH research, we are reminded of the value of putting ourselves out there and taking new risks.

This issue's special feature section (page 18) also looks to the future. When we reflect on how far we have come in the areas of support, advocacy, and research, it becomes easy to see just how far we still have to go. We need to ensure that accessible opportunities are available for those most affected by PH to connect with others who just "get it". We need to continue our collective fight for equal access to all Health Canada-approved treatments. And we need to keep working together towards the day when each individual patient is diagnosed and treated in such a way that allows for a long, healthy life. Whether that occurs in the form of more effective treatments with less side effects—or hopefully even one day a cure—we know we are on the road to a better life for those affected by PH.

Along the way, you will be asked to contribute. Because no matter how hard we work on your behalf, PHA Canada cannot succeed without you. You will be asked to help educate people on what *Life in Purple* looks like during November Awareness Month (page 31). You will be asked to volunteer for committees and special projects so that our work is always informed by your needs and your expertise (www.phacanada.ca/volunteer). And you will be asked to donate or raise funds so that we can ensure a sustainable future for our organization (www.phacanada.ca/donate). It is thanks to your support and engagement—both practical and financial—that PHA Canada embarks on its second decade with confidence and optimism.

Not surprisingly, change has been a consistent theme for PHA Canada over the past 10 years. Each year it has been easy to look back and remark on the progress we have made together. I have no doubt that this will continue to be the case. Like a teenager coming into their own and pushing the boundaries of what is possible, I believe that PHA Canada has found its voice and is not afraid to use it. That voice is made up of all of you. It is an echo of your needs and desires, and of our shared commitment to building a brighter future for you and your family. It is a unified voice, and it only gets stronger with each passing year.

Thank you for 10 PHabulous years—we can't wait to see what the next 10 bring!

A handwritten signature in black ink that reads "Jamie Myrah". The signature is fluid and cursive.

Jamie Myrah
Executive Director, PHA Canada

MEMO: INSIDE PHA CANADA AT A GLANCE

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

PHA Canada Appoints Two New Members to Board of Directors

We are thrilled to welcome Ed Rathonyi and Janette Reyes to the Board of Directors of PHA Canada! Both will bring new skills and perspectives to the group. You can find out more about them below and by visiting www.phacanada.ca/whoarewe.



Ed Rathonyi

Ed hopes to contribute to the empowerment of Canadians affected by PH and give back to the community for the support he and his family have received throughout their journey with PH.



Janette Reyes

Janette is a strong advocate for the integral role that nurses play in the care of PH patients and is incredibly enthusiastic for the opportunity to bring their voice to the Board of Directors.

Ambassador Program Grows!

PHA Canada's team of Ambassadors work to extend the organization's reach across the country and inspire their peers, supporting them to share their stories and raise their voices.

We are welcoming two new Ambassadors to the team! Judith Moatti—from Montreal, Quebec—is a PH patient who leads the Montreal Support Group and works to share her story as a means to raising awareness of this illness. Sonya Collins—from Paradise, Newfoundland & Labrador—is also a patient who, against all odds, helped develop a PH network of PHriends in her region and has advocated for better access to treatment locally and nationally. Both will be seeking to raise awareness of PH and represent our community in their regions.



Judith Moatti



Sonya Collins

New PH Community Resources: Oral Medication Fact Sheets



In collaboration with the Canadian PH Professionals Network (CPHPN), PHA Canada has published a series of oral medication information sheets that cover everything you need to know when prescribed oral medications to treat PH. You can download these by visiting www.phacanada.ca/resources.

PHA Canada works continuously in cooperation with special advisory committees and the community to develop free tools for better understanding and managing life with PH. From fact sheets to tips on the daily management of PH, you can browse through resources by visiting www.phacanada.ca/resources.



ASSOCIATION OF CANADA

2008 - 2018 L'ASSOCIATION D'HYPERTENSION PULMONAIRE DU CANADA

A UNE COMMUN

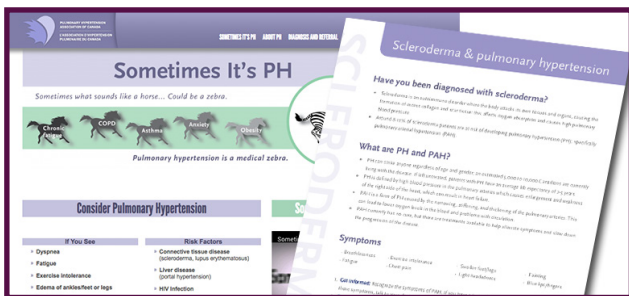


PHA Canada's Board of Directors during AGM in April 2018.
Missing from photo: Carolyn Doyle-Cox, Jennifer Gendron

Early Diagnosis: 'At Risk' Information Sheets

This summer, our temporary Education Campaign Coordinator, Garnet Barrett, developed a series of information sheets for patients with conditions that put them at increased risk of pulmonary hypertension.

We encourage the community to view these by visiting PHA Canada's early diagnosis campaign website:
www.SometimesItsPH.ca.



PHA Canada Departures

We wish to extend our most sincere gratitude to these incredible individuals who have committed their time and expertise over the years to empowering and strengthening the PH community:

- **Carolyn Doyle-Cox:**
Chair, Canadian PH Professionals Network, 2014-2018
- **Teri Kingston:**
Ambassador, 2014-2018
- **Kerry Pierce:**
Pediatrics Committee Member, 2011-2018
- **Sarah Platner:**
Pediatrics Committee Member and Chair, 2011-2018

Inside PHA's 2018 International Conference:

The Pulmonary Hypertension Association's *International PH Conference and Scientific Sessions* took place in Orlando, Florida from June 29th to July 1st. PHA Canada was represented by our Executive Director, Jamie Myrah, along with Ambassadors Beth Slaunwhite & Judith Moatti and Board Directors Nicole Dempsey & Renée Levaque. Several Canadian patients, caregivers, and nurses also made the trip, creating a strong Canadian presence at this year's event. The theme for the conference was "PHinding Your Hope", which is exactly what they left with.

The day before the conference started, seven Canadians attended the *International Leader's Summit*, which included representatives from PH associations from over 30



Canadian participants gathered at PHA Canada meet & greet

countries around the world! The next day, the conference was officially opened with two inspiring keynote addresses by PH patients Nazera Wright and Elvis Medrano. Newly diagnosed with PAH, Nazera Wright shared how connecting with PHA and support from her family helped her to find her new normal. And PHA's 2018 celebrity champion, boxer Elvis Medra-

no, talked about his battle with CTEPH and his journey to getting back in the ring where he now trains six days a week. In the days that followed, participants attended sessions to learn from medical experts, patients, and caregivers, connected with their peers in special support group sessions, learned about new research, and explored a dynamic exhibit hall full of useful demonstrations and fun treats!

You can view a full recap of the conference and access handouts from the sessions by visiting www.phassociation.org/education-programs/conference. ■

Your Community In Action

Community Events

Over the past few months, the Canadian PHamily's calendar has been filled with various awareness and fundraising activities! We are thrilled to share with you highlights and photographs from some of the community events that took place between April and August 2018. Thank you to all of the event organizers, participants, volunteers, donors, and sponsors who contributed to making each event a PHantastic success!

Dolores's Heavenly Heart (April 28th, Winnipeg, MB)

Corrise Proulx and Samantha Roy in Winnipeg (MB) organized a very special awareness and fundraising event for the first time this year. The event honoured their mother, Dolores Rapinchuk, whom they lost to PH in September 2016. Their family and friends came together to celebrate her life, while also raising awareness of pulmonary hypertension. From wearing periwinkle to holding a

special program, the event drew PHenomenal support from the local community and raised over \$1,500 for PHA Canada. We are so grateful to everyone who attended and helped raise funds for the Canadian PH Community, and a very special thank you to Corrise and Samantha for organizing this event.



Some of those attending wore purple to show their support of the PH community.

World PH Day (May 5th, throughout Canada and internationally)

This year marked the 7th Annual World PH Day! Every spring, the global PHamily comes together to celebrate the strength and resiliency of those affected by this disease worldwide, along with developments in PH research and treatment. In Canada, our national *Paint Canada Purple for World PH Day* campaign saw nine monuments illuminated across the country (including the likes of Toronto's CN Tower and Vancouver's Sail of Lights!). There was a

flurry of periwinkle power demonstrated on social media. And—in conjunction with our 10th anniversary—PHA Canada made a very special return to the Vancouver Public Library for an awareness event with local community members, board members, and clinical staff. Thank you to everyone for making this year's *World PH Day* celebrations so special and for continuing to raise awareness of the disease within your local communities.



PHA Canada staff at the Vancouver Public Library raising awareness of PH.

The Zloty family showing their support.

Monuments across Canada were lit up in purple for *Paint Canada Purple!*



Full periwinkle power at this year's *Ajax Run/Walk for PH Research!*

Fifth Annual Ajax Run/Walk for PH Research (June 9th, Ajax, ON)

Now in its fifth consecutive year, we are thrilled to report that the *Annual Run/Walk for PH Research* raised enough money for PHA Canada to fund a new *Mohammed Family PH Research Scholarship!* Thank you Mohammed Family! The event drew 155 participants to Ajax Rotary Park on Saturday, June 9 to run, walk, or

cheer on participants in support of the PH community. The group raised over \$23,000! All of us at PHA Canada want to thank and congratulate everyone who helped make this event such a huge success, and a special thank you to Renae, Joseph, Judy, and Kam Mohammed for organizing their most successful event yet.



All smiles at this year's *Ajax Annual Run/Walk for PH Research!*

Fourth Annual GolPH for PH Tournament (July 20th, Brampton, ON)

It was all sunshine and smiles at this year's *Annual GolPH for PH Tournament!* And not only did the participants take to the greens for a PHabulous time, they also raised \$19,000 for the PH community! We would like to extend our most sincere congratulations and gratitude to the Paulin Family for organizing this important fundraiser in support of PHA Canada's programs and services,

including our PH research scholarship program. Thank you to all the *GolPH for PH* participants, donors, sponsors, and volunteers who contributed to making this year's tournament a hole in one! And we are especially grateful to Unither, Actelion, and Bayer for their generous sponsorship of the event.

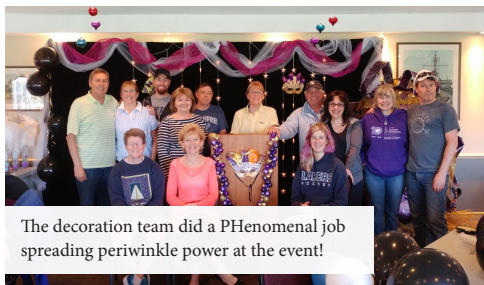


Participants didn't just play an incredible game of GolPH, they also learned about pulmonary hypertension.

Fundraising 101:

Putting on the 2018 Ottawa 'Unmasking PH' Masquerade Ball

Following her PH diagnosis in late 2016, Susan Bailey has continuously worked hard to advocate on behalf of the entire pulmonary hypertension community, raising awareness of the disease all around her and fundraising to help support those affected by this disease. Ottawa's 'Unmasking PH' raised an incredible \$9,000 for the national community this year. Thank you to the entire event committee for their hard work and congratulations on what was a PHenomenal event!



The decoration team did a PHenomenal job spreading periwinkle power at the event!



Susan Bailey with her husband Jim at the event.



The seed for the Ottawa 'Unmasking PH' Masquerade Ball was planted in July of 2017, when a connection in Calgary had the idea that cities across Canada could simultaneously host an event to honour *World PH Day* on May 5th. When I brought this idea to our support group here in Ottawa, they were immediately on board. It is my hope that this article can serve as inspiration to others in the community, so that together we can continue to raise awareness of pulmonary hypertension across Canada.

Our vision for the event was three-pronged: spread awareness about PH, enjoy the event, and ultimately raise some money to benefit patients and families affected by this terrible disease. The first step to any successful fundraising event of this scale is to form an event committee from a group of people who all bring something different to the table: creativity, spreadsheet skills, a decorating vision, fundraising/sponsorship skills, and overall organizational abilities. It is worthwhile noting that people who join a committee like this should expect to work long hours and be dedicated to a common vision. Moreover, the group should not consist of too many patients even though they may be eager to join. The fatigue experienced by a PH patient can sometimes be overwhelming and, as a result, it may be necessary to reduce their load. In my case, for example, the committee asked me to leave the decorating to others and to rest before the evening festivities where I was heavily involved.

Once we had our committee assembled, it was important that we secure a venue early on. I checked out some places and landed on our golf club, Hylands Golf. This was not just a huge relief, it also meant we could get started planning the program for the event. Our focus was on awareness and fundraising. The latter can come in so many shapes and forms, but our group ultimately felt that an auction would be appropriate for the type of event we were planning. In our case, we planned both a live and a silent auction. The first is when a loud auctioneer takes bids from an assembled

group, usually for items that will draw a higher bid value. A silent auction accepts bids from participants in writing for items put on display. So, throughout the summer and fall, we hunted down silent and live auction items in preparation for the big night. This part can sometimes feel overwhelming, but a good place to start is visiting local businesses where you have been before or where they know you. We asked for things like gift baskets, gift cards, and other items we thought our community would want

to bid on. Sometimes we had to return to the same businesses a few times, depending on the amount of money they had available for charitable gifts. It is also a good idea to "think outside of the box"; for example, a neighbour offered to prepare an Indian dinner for our silent auction!

Another thing to consider is sponsorship. Sponsorship is where businesses such as banks, larger private businesses, dentists, and pharmacies provide financial support for your event in exchange for advertising. This was very important to us since they provided "working money" for purchasing decorations, thank you cards, posters, placemats, and event booklets.

We kept our sponsorship levels reasonable with the lowest being \$250 and the highest being \$1,000.

It was our primary mission to highlight and really bring awareness to PH and its impact on patients and their families. The décor for the evening was one of the main tools for achieving this. The 150 attendees were encouraged to wear masks to emphasize the idea that PH is an invisible illness. Around the room there were personal stories from some of the Ottawa support group patients and a tribute frame to commemorate those who had recently passed away from PH. There were also two rolling videos: one with a Masquerade theme and another with photos of the Ottawa support group activities over the years, as well as credits to our major sponsors. Our emcee for the night was a local TV personality, Joel Haslam. He was already very knowledgeable

“It was our primary mission to highlight and really bring awareness to PH and its impact on patients and their families”

about PH from having previously interviewed a couple of patients about living with the disease. He brought a presence and levity to the evening that truly makes a difference on a night like this.

We only had a few key speakers during the evening: my introductory remarks, Dr. Lisa Mielniczuk (Medical Director of the Ottawa PH Clinic), Chris Mainwood, who spoke about being a caregiver, and Nic and Kasia Chartrand who had recently lost their infant daughter to PH. The messages were uplifting and meaningful. Another important aspect of the evening was the surprise presentation of the *Purple Heart Award* to Carolyn Doyle-Cox—the Advanced Practice Nurse (NP) for the PH Unit at the University of Ottawa Heart Institute—for her incredible

service to PH patients and their families all around the country, and particularly in the Ottawa area.

'*Unmasking PH*' was a total success! It went far beyond our expectations and, from having asked several attendees for feedback, I noted that they all indicated that the evening was spectacular and that they would love to return another year. Will we hold this event again? Absolutely! Our hope is to see it succeed once again and to continue to "give back"—particularly to our own community—and to inspire other cities to take part in the concept and put on this event in their region. ■

Contributed by: Susan Bailey, PH Patient, Ottawa, ON

Fondation HTAPQ News

Now in its 11th year, "La Fondation Hypertension Artérielle Pulmonaire Quebec" (Fondation HTAPQ) has approximately 500 members, of whom 150 are PAH patients and 350 are contributing supporters. Here we share with you a short update on highlights from the past six months, contributed by their Vice-President, Dolorès Carrier.



Our PHriends in Quebec joining in on our Anniversary with fun props and smiles!



Stéphanie Théoret, Judith Moatti, and Andréane Mailloux in full force at their event in Saint-Eustache, Quebec.

During our AGM this year, we bid farewell to Michel Anctil who had been our Treasurer since 2007. We also took the opportunity to thank our volunteer of the year, Claire Ruel, for her incredible dedication to our organization.

The Board of Directors also expressed their wish to define our organizational path for the future, so as to ensure the longevity and development of HTAPQ. As such, we have begun developing our strategic plan in collaboration with external consultants. Four main groups have been consulted: current and past members of the Board, PH patients and caregivers, volunteers, and medical professionals. The results of the survey will be announced in the fall of 2018.

Looking ahead, we are planning to host brunch-conferences in various regions

of Quebec in order to encourage patient engagement within the community. This is in line with the Board's desire to respond to the needs of HTAPQ's members.

A number of fundraising events took place these past few months:

- Carol Fournier—whose cousin is living with PH—put on *Show RESPIRE* for the second consecutive year. Famous singers and musicians performed, making the event a great success.
- Andréane Mailloux, a young woman living with PAH, organized a fundraising dinner in her region. She received help from Stéphanie Théoret and Judith Moatti—both also PAH patients. All three, along with Sandy Vachon, presented their personal PH journeys, highlighting the need for

earlier diagnosis, organ donation, mental health, and the importance of advocacy.

- In the spring, our annual cheese sale ("vente de fromage") was in full swing. The cheeses from l'Abbaye St-Benoît-du-Lac were packaged up in Plessisville (QC) and distributed across the province with the help of our amazing volunteers.
- Our growing book sale required a great deal of work from both HTAPQ and volunteers this year. We rented a large tent and sold thousands of discounted books. Thank you to everyone who came for the day! ■

Contributed by: Dolorès Carrier, Vice-President, HTAPQ

Your Stories

The Canadian PH community is made up of many exceptional individuals: patients, caregivers, healthcare providers, researchers, and supporters who individually and collectively refuse to give up in the face of the many challenges brought on by this disease. Every individual in our PHfamily has a unique story to tell and can help make a difference in the lives of Canadians affected by PH. We are happy to feature the stories of exceptional PHighters whose unwavering commitment to the cause, unique journeys, or remarkable accomplishments deserve the spotlight.

My Experience Participating in a Human Stem Cell Trial

Jo-Anne Mainwood is one of the few Canadian PH patients to be taking part in a unique clinical research trial studying the possibility of using human stem cells to treat PAH (see page 22). Jo-Anne quickly became a beloved member of the community after her PH diagnosis by starting a support group in Ottawa. Today, she is still living in Ottawa (ON) with her family, where she continues—through hard determination—to live life to the fullest.



Jo-Anne Mainwood pictured alongside her family.

My PH journey began like many others: I began to notice I felt “off”. I would walk up a flight of stairs and need to catch my breath. Head fog would set in and I’d struggle with following group conversations—simply talking would make me out of breath! I felt like I wasn’t being the parent I wanted to be to my two fantastic children, Graham and Kirsten. My incredible husband, Chris, took care of the household, never complaining when I couldn’t get off the couch. At first, my family doctor diagnosed me as having depression and provided me with meds. Seeing as that did not solve the breathlessness, I was referred to a respirologist; that is when I first met Dr. Chandy. He listened to my concerns, ran tests, and diagnosed me with asthma. In spite of the puffers he prescribed, I continued to struggle with stairs and inclines. At my next appointment, I emphasized my breathing difficulties and how I felt achy and spacey. We made a plan involving a series of tests, and that’s when a VQ scan raised the first of many red flags. The test showed what appeared to be blood clots in my lungs. I was immediately admitted to hospital and injected with blood thinners. After a week in hospital, a CT scan showed I didn’t have blood clots after all. That’s when they sent me for a right heart catheter and the results were in: I had Idiopathic Pulmonary Arterial Hypertension (IPAH).

By this time, the disease had stretched and reached into so many aspects of my life. I was emotionally and physically spent by 5pm on most days, working and living in a “breathing typical” world. And although I am eternally grateful for the amazing support of my family and medical team (Dr. Chandy and Advanced Practice Nurse Carolyn Doyle-Cox), I wanted to connect with others in the community. So I started an Ottawa support group. Due to balancing both family and work, I was incredibly grateful to Charlene and Teri for restarting the group after I needed to let it go.

Our first meeting featured a talk by Dr. Stewart, a stem cell expert. He spoke about the excellent results he was having injecting enhanced stem cells into the pulmonary arteries of mice with PH. His research (called SAPPHERE) was very promising, and I was (of course) instantly interested in getting involved. The purpose of the study was to learn more about the long-term safety and effectiveness of “gene-enhanced” cell therapy for PAH. It investigates whether monocyte cells, enhanced with a small loop of DNA, will repair blood vessels by using a protein called human endothelial nitric oxide synthase (or eNOS). Participants in the study would receive either a placebo or the study product by intravenous injections. The enhanced injected cells travel to the smallest arteries of the lungs, which is the site of the vessel-damage that causes PAH to develop.

The criteria to be part of the study are pretty intense. Participants have to sign a consent form, which states the possible risks of the study (abnormal growth of cells, tumours, embolization, etc.). Despite the risks I didn’t hesitate, I wanted a chance to beat this disease at all costs. I also had to go through numerous tests to make sure I was sick enough, but also well enough to participate. The VQ scan from eight years ago—the one that gave an initial



Jo-Anne (center) pictured at the 2013 National PH Conference with patient & advocate Loretta C. (left) and husband Chris Mainwood (right).

Arm 1	Arm 2	Arm 3
<p>Participants receive 4 monthly doses of placebo during the first 6 months, and 4 doses of cell therapy product during the last 6 months</p>	<p>Participants receive 4 monthly doses of cell therapy product during the first 6 months and 4 monthly doses of placebo during the second 6 months</p>	<p>Participants receive 8 doses of cell therapy over the 12 months</p>

diagnosis of blood clots on my lungs—came back to haunt me. I needed to get an angiogram and a chest x-ray to rule clots out. Once that was done, I was accepted into the trial. The study is a “three armed” trial with eight doses administered to a patient over a 12-month period. Many medical tests are required before and throughout the study. My walk test was just bad enough to qualify me, but ruled out other potential participants. Walking down a straight hospital walkway is very different than walking on stairs or an incline. Add some hills or stairs into any walk, and my results would have been significantly worse.

The first step of the stem cell process is called Apheresis. Basically, I was hooked up to a machine while my monocyte cells were harvested, and then my blood (minus the monocyte cells) was returned to my body. This process went on for about six hours. It felt like being inside an amusement park, like I was vibrating/tingling. This was apparently due to calcium depletion so I was simply given calcium. Two fantastic nurses monitored me: Julia, the study coordinator, and Martha, the Apheresis specialist. I received my first injection about a week after my Apheresis.

Please note that I did not, and still don’t, know whether I was in the group of patients that received the actual product treatment or a placebo. But what I can say is that shortly after the injection, it was as if I already started to feel better. Stairs are my number one nemesis, but after the injections my breathlessness on exertion seemed to decrease. In the past few months, I have done

more activities than I have in years: I even went cross-country skiing! Of course no one is aware of which “arm” of the study I belong to, and this information won’t be revealed for at least three years. But it sure feels like the real thing!

By this time, the disease had stretched and reached into so many aspects of my life. I was emotionally and physically spent by 5pm on most days, working and living in a “breathing typical” world.

PH is a constant “cat and mouse game” to stay ahead of the effects of the disease. But if we—as PH patients—continue to work with researchers in the medical field, perhaps we can together find a cure one day. If the trial I am on can help repair or ease the damage done to the pulmonary arteries, then this cell enhancement therapy needs to become a treatment option as soon as possible. I definitely feel better on this treatment and I hope it will help others living with this disease. ■

Contributed by: Jo-Anne Mainwood, PH patient, Ottawa, ON



The Apheresis procedure.

Nothing About Us, Without Us:

Being Your Best Advocate

Currently living in Halifax (NS), Beth Slaunwhite is a dedicated and motivated advocate for the PH community. After being diagnosed with idiopathic PAH in February 2016, she quickly joined PHA Canada's Ambassador Program and has been working to drive awareness and advocacy in the Atlantic provinces ever since. This year, she was invited to attend the 2018 *Health-e-Voices Conference*: an event centered around how health advocates can use media as a tool to amplify their voice.

Several years ago, I found myself making excuses for being tired doing the simplest things, like carrying groceries or walking up hills. As it progressively got worse, I sought medical advice. At first, they said it was most likely exercise-induced asthma and gave me inhalers that never worked. I knew it wasn't asthma but no one was listening to me. Then came the chest pain and the visits to the ER. They determined everything was normal except the symptoms, and so labelled it stable angina. The last time in, they called it "unstable" since I had chest pain from sitting and talking, and hooked me up to a monitor overnight. I had multiple tests but nothing showed, until a cardiology resident asked me to follow him to another room and noticed my difficulty keeping up. He cancelled the impending stress test and reluctantly requested an echo which, surprisingly, showed pulmonary hypertension.

What have I learned from all of this? That we—as patients—are our own best advocates. After my diagnosis, I found out about PHA Canada and the PH community. One day, after reading *Connections*, I stopped in to my local MLA's office to express the dire need for physicians to be able to decide what medications to prescribe (and to not leave it in the hands of bureaucrats!). I also sought help from a support group in Toronto, 2,000 km away! They welcomed me with open arms and *boy* did I find out how much I could cry—I can't thank Ruth Dolan enough for sticking with me through all of it. And sure enough, I decided to get involved and find other patients in my home province that might be going through what I went through. Shortly after this, I found and applied for a position within PHA Canada's Ambassador Program. It wasn't long until I had an interview, and before I knew it I joined the team!

As one of my first "missions", I had the honour of being one of two representatives of the Canadian PHamily at the *Health-e-Voices Conference*, in Chicago, IL. The purpose of the event was to bring together people affected by, or suffering from, a chronic illness, who are leaders and/or advocates for their communities. I cannot say enough about how humbled I was to be amongst them. Most of them were bloggers, photographers, videographers, and all of them had one thing in common: a burning desire to raise awareness of the disease(s) they represent. There were patients with a myriad of illnesses, be it mental illness, HIV, diabetes, breast cancer, lupus, lymphoma, chronic pain, and many more. I did not personally talk

to all 122 advocates, but I would recognize most of them anywhere. These are my people—I felt at one with these incredible advocates. They use their voices to raise awareness; they shout it from every social media platform they can find. They have a need for their communities to be recognized, funded, and ultimately united. I felt that they were already doing what I wanted to do, and knew I could learn so much from the hurdles they had already overcome.

“These are my people—I felt at one with these incredible advocates. They use their voices to raise awareness; they shout it from every social media platform they can find. They have a need for their communities to be recognized, funded, and ultimately united.”

Up until this point, I had never posted on Instagram nor sent a tweet, so I was pretty nervous. In fact, I felt kind of lost at times, especially in a place where I was surrounded by tech-savvy people with far more experience than me in all things media. But I delved in headfirst and learned a lot! The three-day conference was jam-packed. There were sessions on how to run a podcast, how to make videos to enhance your message, how to better tell your story, how to create good visual elements on your computer, and how to use things like Twitter to effectively get your message out. In the midst of all of this, I also learned a phrase that really struck me: “Nothing About Us, Without Us”. What does it mean? Well... It comes from an old

Latin phrase, and means that no decisions should be made without the involvement of those it affects. It reminded me of the day I went to my local MLA's office back home to advocate for better access-to-treatment. And how wonderful, even though mediums and media have evolved over time, that thoughts and ideas can remain untouched.

As most of my friends know, I have embraced my PH and am determined to reach other PHers so that none of us will feel alone. In Canada, we are spread out over a large geographical area, and many sufferers feel very isolated. My goal as an Ambassador for PHA Canada has been to reach out to other patients and families in the Atlantic Region, and connect them to our community. It is hard enough trying to understand this disease, and no one should have to go through it alone. If you are new to PH and new to a chronic illness, I encourage you to use social media to reach out and find others with this disease, and to get involved in the PH community. If you would like to find out more about *Health-e-Voices*, you can view the entire conference online at www.HealtheVoices.com. ■

Contributed by: Beth Slaunwhite, PH patient & PHA Canada Ambassador, Halifax, NS

Adapting to My New Normal

Interview with a PH Patient



Olga with her son in 2001.

Can you tell us a little about yourself?

My name is Olga; I was originally born and raised in a town in El Salvador. When I was approximately 20-years-old I decided to move to Canada. I initially lived in Vancouver, BC, until 1997. Then I moved to Bellingham, Washington (USA). That is where I married my husband. As much as we liked it there, we ended up moving back to Vancouver pretty soon after that due to my illness. We have a son together who is 18-years-old this year.

How did you first find out you have PH?

It wasn't quite so simple. During my pregnancy in 2000, I began to feel a lot of pain in my joints and my hips especially. Even after I'd given birth, the pain was getting worse. The doctor kept saying the pregnancy had affected my immune system. The clinic went as far as advising me to have another child, as this might somehow balance my system out. But by this point my muscles were very weak and hurting day and night.

How long did it take until you were given an actual diagnosis?

There wasn't just one, they diagnosed me a couple of times before we got to the correct ones. After almost two years of tests and visits to the doctor, they first told me I had rheumatism. I was concerned and told the doctor I didn't believe this; I know people with rheumatic disorders

and this wasn't it. Later that year, during a trip home to El Salvador, I met with a relative who worked in the medical field. He asked me to send him all of my test results from the last couple of years. A couple of days later, he called and said I have lupus. My doctor didn't take long to confirm this. My symptoms kept getting worse and treatment wasn't working, so in 2010 I was finally referred to a PH doctor who diagnosed me as having lupus-associated PH. I was still not responding to initial treatments, and in 2012—when it was getting worse—they gave me a choice: I could either get on a pump or have a double lung transplant. It took me a year to agree to use the pump.

“But what makes it worse are people who think they can pass judgment when they don't know anything about you.”

How come it took so long to make that decision?

It was a scary decision because you carry this thing 24 hours per day, everyday, for the rest of your life. But they insisted that the pump was safe and that there were patients who had lived with it for up to 17 years, so that's what I did.

Now that you've received an accurate diagnosis, how are you coping with your “new normal”?

There are limitations: I want to go hiking but I can't; I get worse when it's hot outside; I get easily fatigued. My husband and son love to go swimming, but obviously I can't do that either. So it's definitely tough.

But what makes it worse are people who think they can pass judgment when they

don't know anything about you. In winter-time, the temperature makes me stiff and my joints hurt a lot more than they would any other time of the year. So I often use handicapped parking to avoid having to walk across parking lots to get to the store. People get so mad when they see me do that, because you might not tell that I'm sick from looking at me. So I'll show them the sign to prove that I'm not doing anything wrong. But by that time, they'll have shouted pretty awful things at you.

What would you tell someone who is newly diagnosed and faced with this kind of behaviour?

One of the things I would say to someone who is newly diagnosed is to accept the situation. For me, that was the hard part. You are sick, you won't be the same person you were before, and as long as you don't accept that then you will have a problem. And it takes a while! For me it took years to really say, “Okay, I have to accept this”.

Last year was my first time seeing a huge baseball game in Seattle—you see, I used to be so afraid of crowds. But it was an amazing experience and I'm happy that I've finally started to feel comfortable enough to do things outside my comfort zone. It's starting to feel like things are going to be okay. ■

Contributed by: Olga Leal, PH Patient, Vancouver, BC



Olga overcame her fear of crowds and attended a baseball game this summer with her son!

What Does PHA Canada Mean to You?

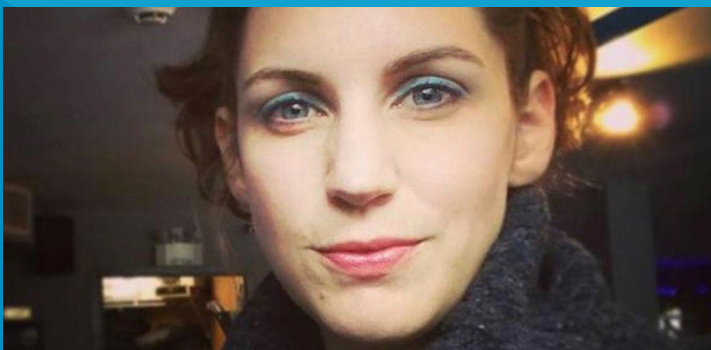
PHA Canada's mission is to empower the Canadian pulmonary hypertension community through support, education, advocacy, awareness, and research. To fulfill this mission and achieve our vision of a better life for all Canadians affected by PH, we offer a variety of programs and services to the PH community. Acting as a resource hub, our organization empowers community members to become educated about PH, as well as to support one another and advocate on behalf of the PH community.

In 2018, PHA Canada celebrates its 10th anniversary. As part of this milestone, we have asked our community how our organization has impacted their lives. Here's a selection of quotes from members of the Canadian PHamily who reflect on what PHA Canada means to them.



“PHA Canada, to me, means unity and not feeling alone in a world where you look healthy on the outside, but feel sick on the inside. The only people that actually “get it” are others that walk in your shoes. Being from a small community and not knowing anyone in my part of the province who is also living with PH is hard. But now I’ve met people that I’m lucky to call my PHriends all throughout Canada. I hope PHA Canada continues connecting us and educating everyone on this disease that has its hold on our lives. Periwinkle love!”

— Jodie Ashini, PH patient, North West River (NL)



“My road to diagnosis has been a long one filled with misinterpretation, misunderstanding, and misguided medical professionals who are trained to look for everything BUT a zebra. PHA Canada understands this struggle and has created resources to help the people in my life to better understand me.”

—Jesse Nicole, PH patient, Windsor (ON)



“What PHA Canada means to me is that the work my family started is continuing and it’s a happy reminder that PH patients in Canada now have so much more support, resources, and options than they did when my sister Sherry was diagnosed in the 1990’s.”

—Philip Paroian, PH supporter, Olympia (USA)



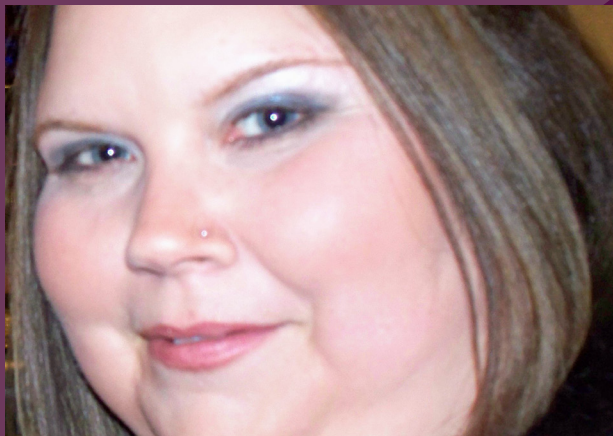
When we received Quentin’s diagnosis, I can honestly say that PHA Canada was our saving grace in a time where we had more questions than answers. We attended the PH conference in Montreal and we found our PHamily! We received the answers we were looking for, we had hope for the future after hearing the stories from the PHighters and their caregivers. PHA Canada gives a sense of community, providing support, encouragement, and information. We will forever be grateful to have such an amazing support system and to have a great group of people who help keep his memory alive through fundraising and education.

— Jolene Mosiondz, PH caregiver, Thompson (MB)



PHA Canada is our family that has been our support and life line on this incredible journey to a successful double lung transplant...April 17, 2018. Thank you for being here for us.

—Blenkinsop Family, Toronto (ON)



When I was diagnosed in May 2017, I felt lost and alone. Thankfully I was introduced to PHA Canada. Through them I found support, understanding, and friendship. They educated me and pointed me towards others that can relate. Now, with PHA Canada as my companion, I know I can handle whatever this condition throws at me.

—Jennifer Adams, PH patient, Mount Pearl (NL)



To me, being newly diagnosed, PHA Canada represents a safe, supportive environment where I can share my personal journey, ask questions, and access resources. It was a “game changer”. I was able to connect with persons like me and hear encouraging words like “you got this” and read others’ stories and journeys.

PHA Canada is a community rich in resources, advocacy, and professional experts. Most of all, it is people like me and people fighting for me.

—Pat Fleming, PH patient, Dashwood (ON)



*Sharon Proudfoot,
PHA Canada
Founding Member*

Dear PHriends,

Wow, Fall 2018! This marks 20 years since I first saw the words pulmonary hypertension scribbled across an echo requisition. In 1998, after spending three years searching for a diagnosis, I finally met with a cardiologist. He listened to me, did an EKG, and said “I think I know what’s wrong with you, but let’s wait until we do the testing”. Being the tenacious person that I am, I took the liberty of looking at the echo requisition when the receptionist stepped away from her desk. That’s when I saw it: “suspected PH??”. I promptly went home and searched for it online. That night, I sat in front of the computer devastated—I knew immediately that I had finally found it.

In the years that followed, I got to meet and befriend other patients in the Calgary PH community. Most of them were much more ill than I was—I had been lucky that I was a responder to nitric oxide, so my pressures responded very well to treatment. That is when I decided I wanted to commit some of my energy to PH—energy that most of my friends did not have—to improve conditions for those living with or affected by this disease. I felt from the very start that my preference was to try and pool resources across Canada for a larger, stronger, and more unified voice. In 2006, I met a group of like-minded Canadians at the PHA conference in Minneapolis, USA. The seed was planted and shortly after that, a small group of us became the Founding Members of PHA Canada: an association focused on empowering and uniting Canadians affected by PH through support, education, advocacy, awareness, and research.

Community support needed to be a vital element for this new organization. My phone number was on published materials and on the website as the point of contact for Alberta. Newly diagnosed patients would call just so they could talk to someone who had experi-

PHA Canada Anniversary Feature

Part II of II:

Celebrating 10 Years

ence with the disease, someone who was living well with it, and someone who could understand some of what they were going through. PH often leaves you feeling very raw and exposed, and not strong enough to put up with a well-meaning neighbor telling you that you should try vitamins or herbal remedies. There is real relief in talking directly or online with people who just “get it”. People who can just cut through the pleasantries and ask what is on your mind. I am very fortunate to currently have that with our very low-key PH coffee group in Vernon, BC. I haven’t known any of these ladies for very long, but we just “get each other”.

Advocacy was also one of the pillars central to the creation of our association. For me, PH advocacy has always been focused on equal and timely access to medication for everyone in Canada. My first foray into PH advocacy was writing letters to Alberta Health asking for the approval of Tracleer® (bosentan) in 2001. Given that each province has decision-making authority for the funding of PH treatments, there needs to be constant vigilance. It takes work and time, but the force of our united voices demanding that PH patients have access to the same medications—no matter what part of the country they happen to be living in—is having an impact (see page 24)!

Looking 10 years ahead, it would of course be wonderful for research to have moved in on a cure. It is important to remember that this is not impossible, but it will require more than patience: it will also require involvement. Patients are invaluable to the process and are in a unique position to help test treatments that could one day help cure those living with this disease. Furthermore, I would like to see progress in communication between PH clinics. Having had the disease for almost 20 years, I have

lived in two different provinces and been to three PH clinics. I can see that they are all run quite differently, with each having their pros and cons from a patient perspective. I think it would be great if there were more standardization of care across clinics and a “best practice model” developed by the clinics—preferably with patient input. I would also like to see transplant be considered as part of the treatment strategy. As someone who has only been on the sidelines, it appears that PH patients often don’t get considered for transplant until, quite frankly, they are bad candidates. I would like to see more aggressive, earlier recognition and initiation of transplantation as a therapy option when the medical team can see that all roads are leading in that direction.

But perhaps most importantly, I would like to continue to see a united community that has access to the kind of immediate support and information that we as patients and caregivers need. And while we have come an incredibly long way in growing as a PHamily, we should never forget the importance of supporting one another throughout our PH journeys.

Regards,



Sharon Proudfoot
Founding Member
PHA Canada

Celebrating from Coast to Coast to Coast

In honour of our 10th Anniversary this year, we decided that—since you could not all come to the party—we would bring the party to you. Thanks to the help of an amazing team of volunteers in Vancouver, BC (picture 13) over fifty anniversary kits were packed and shipped off to all corners of the country. Each of them contained a number of awareness materials and resources, and all the things you might need to kick off your own local anniversary party: a banner, some party hats, photo props, and more. We even included an anniversary themed quiz to test your knowledge of PHA Canada's history: "The PHA Canada logo used to be red & blue. What did each side-by-side colour represent?"

We would like to express our most sincere gratitude to local support groups across Canada and their members for celebrating this special occasion with us. Thank you to the Vancouver Island Support Group and the BC Southern Interior PH Support Group who organized special lunch meetings with their members (pictures 4 and 5). We were thrilled to receive photos from the Ottawa Support Group, proudly holding our birthday banner (picture 8). The London Support Group's event drew an incredible 50 attendees (picture 1); thank you to the organizers for their amazing work putting together decorations and a program for the event. In a first for Nova Scotia, PHA Canada Ambassador Beth Slaunwhite hosted our Executive Director, Jamie Myrah, along with members of the community, for a special anniversary luncheon in Halifax (picture 12).

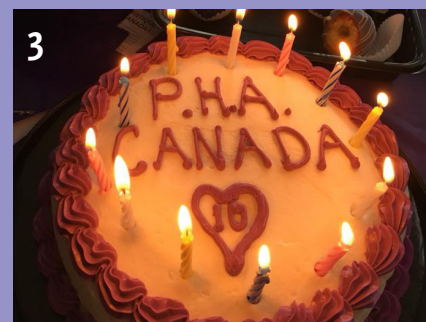
Thank you also to all of the patients,

caregivers, and supporters who ordered a kit and celebrated in their local communities. Kits were sent all the way up north to Kugluktuk, NU, where PH patient Millie Kuliktana had the banner up during a local town parade; to Paradise, NL, where PHA Canada Ambassador Sonya Collins hosted an event with special guest Azlan (picture 11 & 16) and local PHriends; all the way west, where the Vancouver PH Clinic helped welcome local patients for an anniversary get-together (picture 15); and countless places in between, from Toronto (picture 10 and 14) to the Stollery Children's Hospital in Edmonton (picture 9). Throughout the month of April, our anniversary was celebrated in style from coast to coast to coast!

You can see more photos and take part in our anniversary by visiting www.phacanada10.com.

There is incredible strength in unity, something that the Canadian PH community has proven to be true beyond expectations over the past 10 years. So to all those that joined in on the celebration these past few months: thank you for taking the time to support one another—and us—as we continue our work empowering the Canadian PH community for many years to come! ■

Contributed by: Michaël Robach,
Communications & Engagement
Coordinator, PHA Canada





Left to right, top to bottom: (1) London Support Group event; (2) PHA Canada Board of Directors & Staff; (4) BC Southern Interior Support Group; (5) Vancouver Island Support Group; (6) Corporate Committee for PHA Canada and Board Directors Dr. Sanjay Mehta and Roberta Massender; (7) HTAPQ Board of Directors; (8) Ottawa PH Support Group; (9) Janette Reyes (NP) and colleagues at SickKids Hospital; (10) East Side Toronto Anniversary lunch; (11) PH supporter Azlan; (12) Anniversary lunch hosted in Nova Scotia by PHA Canada Ambassador Beth Slaunwhite; (13) Volunteers packing anniversary kits at the PHA Canada office in Vancouver, BC; (14) Ruth Dolan, Loretta C., and Joan Paulin celebrating our anniversary in Toronto, ON; (15) Vancouver PH Clinic Anniversary Party.

The Future of PH Therapies: Zooming In on Canadian PH Research

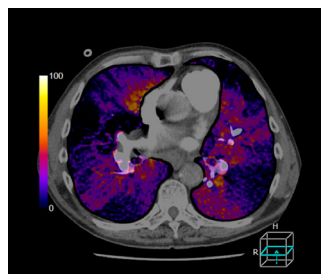
During the past 10 years, PH medical research has evolved immensely throughout the world. Medical professionals are delving deeper into the science and uncovering new information every day. And while this has taken place internationally, we are incredibly fortunate to have leading research and clinical trials taking place right here in Canada.

A clinical trial is a study that is done to decide if a medical treatment is safe and effective for humans. Successful clinical trials can lead to the approval of medical treatments for use by the public. Thus, clinical trials are incredibly important for developing and improving PH treatments. By participating in clinical trials you can directly impact the PH community in a profoundly positive way. Thanks to volunteers from the PH community, doctors have been able to perform many clinical trials for PH in Canada. Here are three examples of research projects that are currently taking place.

CTEPH Research in Canada

*Submitted by Dr. Marc de Perrot and Laura Donahoe,
Division of Thoracic Surgery, Toronto General Hospital*

Chronic thromboembolic pulmonary hypertension (CTEPH) is an increasingly recognized condition that occurs when the body does not clear blood clots in the pulmonary arteries (PAs) of the lungs. This results in chronic scar tissue that causes high blood pressure in the lungs and right-sided heart failure. The standard of care treatment for CTEPH is a surgery called pulmonary thromboendarterectomy (PTE), which cures the disease in a large number of patients through removal of the residual scar tissue in the PAs. One of the main obstacles to management of patients with CTEPH has been making the diagnosis, as the disease is often missed or underestimated on CT scans. Current research focuses on better ways to image the PAs in order to identify the blockages from the scar tissue in the lungs. One promising new technique is to image the perfusion of the lungs by CT scan (image below).



Historically, only patients whose disease was in the main PAs or the first-branches off of the main PAs would be considered for surgery. But advances in the Toronto CTEPH program have demonstrated that surgical management of patients with disease farther out in the lungs, in the second- and third-

branches of the PAs, is also successful. Also, for patients whose disease is still too far out in the lungs for surgery, we have started a Balloon Pulmonary Angioplasty (BPA) program. The idea behind BPA is that the blockages in the PAs caused by the scar tissue in the small lung vessels are stretched to open up the arteries, improve blood flow, and decrease the blood pressure in the lungs.

As the medical community has also become more aware of the phenomenon of CTEPH, residual blood clots in the lungs are being identified in patients with shortness of breath but without PH. The Toronto CTEPH program now offers surgery to those selected patients and continues to study how they do both with and without surgical management. Specific investigations are being performed to assess the impact of these residual blood clots on the functional performance during exercise. We are also conducting research in the thoracic laboratory on the surgical specimens to understand the mechanisms that lead to PH with the hope that we will eventually be able to screen patients at risk of developing PH and diagnose the disease before the development of heart failure.

Finally, educating surgeons in CTEPH surgery is a focus of current research, as PTE is a very difficult surgery to learn and teach. The Toronto CTEPH program is currently undertaking video recordings of PTEs to help educate Canadian and international visitors coming to our center, and has developed a PTE simulator for practice outside of the operating room. ■

SAPPHIRE: Stem-cell Research in PH patients

*Submitted by Dr. Duncan Stewart,
Ottawa Hospital Research Institute*

Is it possible to grow new blood vessels in the lungs to replace the ones that are damaged or blocked? That is the question a new clinical trial for pulmonary arterial hypertension (PAH) is trying to answer.

SAPPHIRE (which stands for “The study of Angiogenic cell therapy for Progressive Pulmonary Hypertension: Intervention with repeat dosing of eNOS enhanced EPCs”) is a trial that will test whether gene-enhanced endothelial progenitor cells (EPCs) therapy is effective and safe for the treatment of severe PAH. This experimental therapy starts with collecting a patient’s own white blood cells, which are sorted into different components. One component is grown in the laboratory under special conditions to obtain cells called endothelial progenitor cells (EPCs). These cells are important in the repair and regrowth of blood vessels; however, to enhance their regenerative activity, they are then genetically engineered to produce greater amounts of nitric oxide, which enlarges blood vessels and stimulates their repair. These gene-enhanced cells are then injected directly into the same patient through a simple intravenous injection to be carried to the lungs.

A previous phase I trial of this therapy (the PHACeT trial) in seven patients showed that the delivery of gene-enhanced cells was well tolerated. Although it was not designed to rigorously assess benefits of the therapy, participants showed improved breathing, exercise ability, and quality of life.

SAPPHIRE is a phase II randomized, controlled, and double-blinded trial that will test whether gene-enhanced EPC therapy is effective and safe for the treatment of severe PAH. It has an innovative design that will allow all participants to receive gene-enhanced cell therapy at some point during the study period. To do this, participants will be randomized to one of three treatment arms (see page 12 for more details). It is anticipated that 45 PAH participants will be enrolled in up to nine participating sites across Canada. As this therapy is experimental, it is impossible to say if patients who receive the therapy will benefit. Further information can be found at www.clinicaltrials.gov. ■

PARP1: PAH is not a cancer, but anti-cancer therapy may be effective

Submitted by Dr. Steeve Provencher, Centre de recherche de l’Institut universitaire de cardiologie et de pneumologie de Québec

Cancer cells are characterized by sustained survival, despite living in stressful environments that damage the cells. They commonly acquire a “super system” to repair cell damage, enabling them to survive almost indefinitely. This appears to be driven by an enzyme called “PARP1”. Recently, researchers have developed newer treatments that specifically inhibit this “super system”. These treatments—blocking the enzyme PARP1 and consequently the survival of cancer cells—were shown to be quite effective in treating certain types of cancer, even when standard chemotherapy was no longer effective. And on top of that, these pills had minimal side effects in humans. These are now approved for the treatment of certain cancers in both Canada and the USA.

Pulmonary arterial hypertension (PAH) is characterized by the progressive narrowing of the pulmonary vessels, leading to persistent increases in pulmonary blood pressure. In normal circumstances, there should be a regular turnover of cell death with replacement by new cells. This turnover should be increased because of the stressful environment caused by the increased pressure within the blood vessels. Canadian researchers documented that PAH cells exhibit the same behaviour as cancer cells, enabling them to survive almost indefinitely and that PARP1 is mostly responsible for excessive cell survival in PAH, which contributes to the progression of PAH. The blockade of the enzyme PARP1 was not only sufficient to stop the abnormal growth of the

cells from human PAH lungs, it also markedly improved the disease in animals with pulmonary hypertension.

Given the limited side-effects of this newly available drug, the time appeared right to explore whether blocking PARP1 could be safe and effective in treating patients with PAH. While researchers would like to make new drugs rapidly available for patients, they also want to make sure it is effective and safe for them. Therefore, a pilot study will be initiated shortly in Quebec City (first patients enrolled expected this Fall), likely followed by further studies if initial results are positive. Initial results are expected by the end of 2019. ■

“PH is a constant “cat and mouse game” to stay ahead of the effects of the disease. But if we—as PH patients—continue to work with researchers in the medical field, perhaps we can together find a cure one day.”

—PH patient participating in SAPPHIRE trial

Together in the PHight:

Advocating for a Better Life

One of the most vital components to PHA Canada's purpose has always been to advocate on behalf of the PH community. To "advocate" can imply many things: it can evoke the process of enabling people to express their views and concerns; it can mean assisting someone in gaining access to information or services; or it can involve championing a particular cause, policy, or decision. But in all cases advocacy gives voice to those who otherwise may go unheard.

Throughout our 10-year history, giving voice to the PH community has always been at the heart of PHA Canada's existence. In May 2008, only a month after the official founding of PHA Canada, MP Don Bell (North Vancouver, BC)—father of Founding Member & President, Darren Bell—introduced a private members motion (M426) in the House of Commons in support of PH and other rare disorders. The motion called on the government to "respond specifically to the challenges faced by Canadians with rare diseases and disorders" and received the support of all parties (except the Bloc Quebecois). Spearheaded by the Canadian Organization for Rare Disorders (CORD)—an umbrella organization in which PHA Canada remains a member today—the motion led to the development of a Canadian Rare Disease Strategy, which CORD officially unveiled in 2015.

Meanwhile, by the end of 2008, the fight for access to optimal treatment was erupting in Ontario. The government had made the decision to limit patients to one PH-targeting therapy at a time, which meant those on combination therapy would have their treatment reduced to one drug. PHA Canada—led by the Toronto Chapter—rallied the community to respond. Complaints to the Ombudsman's Office, meetings with elected officials, media attention, and direct engagement with the Ontario Ministry of Health and Long-Term Care led to a review of the restrictions, which were removed by the Committee to Evaluate Drugs in 2009. However, challenges to the Ontario Provincial Drug Plan would continue in the coming years, as concerns about the criteria for accessing combination therapy persisted.

By 2014 PHA Canada was in need of more resources to help raise awareness and educate decision makers. The Ambassador Program was initiated, and 10 incredible volunteers stepped up for a two-year commitment to represent PHA Canada in communities across the country. PHA Canada's *Burden of Illness*

Report—based on a survey of Canadian patients and caregivers in 2013—was also released. Board, staff, and Ambassadors worked together to engage government officials, writing letters and holding meetings to ensure that decision makers understood the needs of patients and their families.

Efforts ramped up even more in 2015 when the Canadian Agency for Drugs and Technologies in Health (CADTH) released a report recommending a "stepped approach" to the initial treatment of pulmonary arterial hypertension (PAH). PHA Canada launched a campaign advocating that PAH treatment decisions be made by PH expert physicians and not government committees. A letter-writing campaign, along with face-to-face meetings in BC, Saskatchewan, and Ontario, put pressure on the provinces to reject CADTH's recommendations and not restrict the ability of physicians to exercise their individual clinical judgement. PHA Canada also made submissions directly to CADTH, to represent the views of the PH community and ultimately to express disappointment with the agency's conclusions.

During this time, the fight for access to combination therapy was becoming increasingly relevant as new oral PAH therapies entered the market. Along the way, PHA Canada consulted with the PH community on the value of these new treatments, making submissions on their behalf to the Common Drug Review for macitentan (2014), riociguat (2015), and selexipag (2016). In December 2015, PH advocates held an "Advocacy Day" at Queen's Park in Toronto (ON). Patients, caregivers, and medical providers spoke with MPs and the media to tell their stories and discuss the importance of access to new treatments for PAH.

Just weeks later, the panCanadian Pharmaceutical Alliance (pCPA)—who is responsible for negotiating drug prices on behalf of the public drug plans—announced they were closing negotiations for macitentan without coming to a deal with the manufacturer. As a result, Quebec—who conducted its own separate analysis and negotiations—became the only province to provide public funding for macitentan. As we entered 2016, we began to worry that a trend was in sight: a positive recommendation from the Common Drug Review would not guarantee access to patients if pCPA pricing negotiations weren't successful. In January 2016 we responded by launching the *Take Action PAH* campaign (in partnership with Scleroderma Canada), which provides tools to

“PHA Canada continues to work with advocates throughout the country who want to help make the needs of PH patients visible to those who can make a real difference to the quality of care and treatment they receive.”



Don Bell receiving a CORD award in 2017 for his work advocating for rare disease communities.



Dr. Sanjay Mehta, Ruth Dolan, and Nicole Dempsey representing the PH community during the 2015 Advocacy Day at Queen's Park.

Canadian government decision maker

- ✗ Does not know about pulmonary hypertension
- ✗ Does not know you
- ✗ Cares about reducing costs

Canadian PH expert physician

- ✓ Specializes in the treatment of pulmonary hypertension
- ✓ Knows you and your disease personally
- ✓ Cares about your wellbeing

Who do you want making decisions about your PAH treatments?



PH advocates Joan Paulin & Martha Barale (front) along with PHA Canada Executive Director Jamie Myrah (right).

the PH community to help advocate for public access to all Health Canada-approved treatments for PAH. As selexipag entered pCPA negotiations in 2017, PHA Canada led the efforts to encourage those negotiating to make a deal on selexipag and ensure access to patients in need through public funding.

In 2017, PHA Canada also returned to working with the Ontario Ministry of Health and Long-Term Care on their review of the criteria for providing PAH treatments. Written submissions and expert presentations to the Committee to Evaluate Drugs helped contribute to updated criteria that include expanded access to combination therapy. This process also allowed for continued engagement on the need to provide funding for new oral treatment options, and the importance of patient and physician choice in order to achieve optimal treatment.

This year, we saw the successful completion of pricing negotiations for selexipag and the majority of provinces across the country add the drug to their formularies. Unfortunately not all provinces have done so, and of course other therapies continue to be out of reach to patients without private drug coverage. PHA Canada continues to work with advocates throughout the country who want to help make the needs of PH patients visible to those who can make a real difference to the quality of care and treatment they receive. We continue to engage with governments through letters, official submissions, consultations, and meetings on budget priorities, changes to legislation, and issues concerning access to treatment.

For 10 years, PHA Canada has been raising the voice of the Canadian PH community. We have worked diligently to ensure that your stories are heard and your needs are met. We can't always speak for you, but we will provide the tools and support you need to be empowered to speak up. Of course, there will always be more work to do. But the unity of our community gives us the strength and power to effect real change. Join us, as together we continue our fight for a better life for all Canadians affected by PH. ■

Contributed by: Jamie Myrah, Executive Director, PHA Canada

A Physician's Perspective on the Future of PH



*Dr. Sanjay Mehta,
PHA Canada
Founding Member*

Dear PHriends,

I first learned about PH as a medical resident in 1990, looking after a patient who had PH and resulting right-sided heart failure without a clear reason. This led to a research project and publication of my first research paper in 1991. Since then, I have had the great privilege and honour of caring for PH patients for nearly 30 years. During this time, the PH medical communities in Canada and around the world have made great progress. As Yogi Berra once said, “the future ain’t what it used to be”. For PH patients, it certainly isn’t! The future is much brighter than it has ever been.

Ongoing research into the science, diagnosis, and treatment of PH has dramatically changed the daily lives and future of most PH patients. There is no doubt that someone diagnosed with PH today has access to much better care than a patient diagnosed 25 years ago. Canadian PH patients are now assessed and treated in 22 expert PH centres across the country. Moreover, PH patients currently benefit from 10 different effective PH-targeted medications that have been approved by Health Canada over the past 20 years. This has been an unbelievable pace for research and drug development, unparalleled in any other area of medicine. As a result, PH patients are living longer and better lives than ever. For example, average survival was only 2-3 years before PH-targeted medications first became available in 1997. For patients diagnosed with PH today, many will live 10 years and possibly much longer, as survival continues to increase every year.

PH is also an illness that seriously impairs the ability of patients to function in everyday life, to fully participate in life, and to enjoy a good quality of life. As such, improving the health-related quality of life in each patient has become a major focus in the care of PH patients. Available PH-targeted medications can improve quality of life, by reducing the frequency and severity of symptoms, and improving patients’ ability to be physically active. Importantly, effective treatment of PH can keep patients more stable over time, such that they don’t progressively worsen, get hospitalized, or need more aggressive treatment approaches like lung transplantation.


What does the future hold for PH patients and their caregivers? From a medical perspective, I fully expect new treatment approaches, especially “biologic” therapies similar to other fields such as for the treatment of cancers and rheumatic diseases like lupus. Future therapies will be better targeted at the increasingly complex science of the disease. This may include treatments to prevent the death of endothelial cells, the cells that line the inside of the pulmonary arteries (that supply blood to the lungs), the blood vessels that are damaged in PH. Another target will be the excessive growth of cells, including these same endothelial cells, as well as the muscle cells in the walls of the pulmonary arteries.

Importantly, most current therapies are not specifically chosen for individual patients; most PH patients are treated with the same medications. PH doctors today have very little information on which treatments might work best in specific patients. The future will see more of a “Precision Medicine” approach to the treatment of many conditions, including PH. Treatment will be tailored to each individual patient’s genetics, background, and environment. This will help PH doctors treat each patient more appropriately as an individual, with the optimal medication, while avoiding others that may not be as effective or cause more side effects. It should also be noted that we have thus far focused on the care of a narrow spectrum of PH patients, specifically PAH and CTEPH. There is a much larger group of patients who suffer from PH due to heart and lung disease that will increasingly ask for and deserve our help in the future.

A goal for many of our patients, volunteers, doctors, and researchers is finding a cure for PH. It is clear to all of us that none of our current treatment options can cure PH. In fact, most patients on current PH-targeted medications will eventually experience worsening of their PH at some point. I too would of course be thrilled to one day have a cure I could prescribe to patients. There is the hope that such a treatment approach may already exist! The very exciting Canadian research into cell-based gene therapy in the SAPPHIRE Trial (see page 24) may be able to dramatically improve and potentially even reverse PH in some patients, as this therapy has already shown in animal studies.

I am personally uncertain if a cure for PH is currently possible. This is because PH has most likely been present for several years before patients experience symptoms and get medical attention, such that most of the damage to the pulmonary arteries will not be easily reversed. However, I still have great hope for the future of PH patients. I believe new treatments will further improve patients’ daily wellbeing and quality of life, and continue to improve long-term survival. I clearly see a day in my lifetime when a PH patient is diagnosed and treated, such that they function and live normally. They don’t have any symptoms of PH or even have to think about PH on a daily basis, because it has become a chronic condition that they live with, rather than suffer from.

This focus on living well with PH has been the vision of PHA Canada since its founding. Based on our first 10 years, the PHuture of PHA Canada holds great promise! We are committed to our work of supporting, educating, and advocating on behalf of PH patients and their caregivers across Canada. I would like to see, and will continue to work towards, a day when we connect with each and every Canadian affected by PH, inviting them to join us as part of a united national community of support and understanding.



Sanjay Mehta, MD, FRCPC, FCCP
Director, Southwest Ontario PH Clinic
LHSC—Victoria Hospital, London, ON
Chair, Board of Directors, PHA Canada

A National Registry for Canadian PH Patients

For close to a decade, a special taskforce of Canadian PH medical professionals has been working behind-the-scenes on developing a National PH Registry for Canada. And now after testing it out at the Vancouver PH Clinic since January 2017, they have begun the process of recruiting clinics from across Canada to adopt the system.

We had the pleasure of interviewing two members of the committee who are helping to lead the project. Lena Legkaia is the project manager; her focus is to facilitate and coordinate the national implementation of this new clinical database system in PH clinics across the country. Lisa Lee, nurse practitioner (NP) at the Vancouver PH clinic, offers her first-hand expertise in the treatment of PH patients and the importance of such a registry.

To start us off, could you tell us in simple terms what a registry is?

Lena: Registries are very common tools that we see in everyday life. A registry is simply put a program where you store data. They're very useful in the health sector because they allow us to better understand overarching trends and make educated decisions on how to improve or adjust the way patients get treated.

Lisa: Countrywide registries are not a new concept, but this is a first time an effort to implement a Canada-wide PH registry has been made. Health registries are actually very common in the world of pulmonary hypertension and rare diseases in general, because not a lot is known about people who have rare diseases.

What are you hoping to achieve?

Lena: The overarching goal is to have patients' data entered into a database so that down the road, researchers can go—with approval, of course—look at the data and figure out where we need improvement, what is working, and what isn't. We can assume that PH patients in Canada are the same as in the USA and EU (European Union), but we don't actually know. For example, because different provinces have different drugs available for treatment, are patients in certain provinces being treated as well as they are in other areas of the country?

Lisa: Primarily we will look at mortality—so, do patients who have PH in Canada live as long as patients in other countries? There are a lot of other things we look at: a lot of the big drivers right now in the world of registries and PH is looking at risk, so how can we tell that you're at risk or that your PH is at risk

of progressing? Well, they take all this information on all these patients and they see what kind of patients do well and live for a very long time, and then what are the kinds of patients that do poorly. And what are those indicators that are letting us know that a patient might get worse over time, so that we know actually when we can intervene earlier in terms of treatments or different therapies for their pulmonary hypertension.

I imagine privacy must have been a pretty big topic when planning for this project?

Lisa: That was definitely one of the major concerns; it's one of the things that we spent a lot of time working on. There's a lot of oversight from both health authorities and the government in terms of making sure this data can't accidentally get out into the general public. No one will get access to the patients' individual data, it will be amalgamated—kind of de-identified—information. It won't be, you know: "Mr. Bob has this kind of PH, and he tried this medication, and got better". It will be shown like: "We have 750 patients who have pulmonary hypertension in the clinic. These are the types of PH that they had. These are the types of patients that did well. These are the types that did poorly".

Lena: Clinics are using a coded master list where the software generates a random number for each patient that is in the database. And then that number can be linked to their name, but that is stored at the clinic: nobody gets to see it but the physicians and the nurses. So the computer program doesn't actually contain any personal identifying information, only a number and the corresponding health history.

“Countrywide registries are not a new concept, but this is a first time an effort to implement a Canada-wide PH registry has been made.”



Lisa Lee (NP) and Lena Legkaia at the Vancouver PH Clinic.

Do patients have the option of opting out if they don't want their information included?

Lena: Yes, patients can always opt-out. It's a requirement that the clinics get a signed consent from patients when entering their data into this new registry.

Lisa: Interestingly though, we haven't had any push back from patients. We're having our unit clerks collect the consent forms upfront so it's not like they've had to do this big in-depth discussion. Patients are mostly like "oh, you're collecting information to improve PH research? I'm happy to sign up" The information is safe so there really hasn't been an issue.

How does this affect the clinics?

Lisa: If you can imagine, we have 700 patients in our clinic: the amount of resources and infrastructure to coordinate this both on an individual clinic basis is already a lot, and then getting together very busy PH clinics on top of that. We have to figure out which software you're using, submit ethics and research applications, all this kind of stuff. But ultimately, really, what the big change is going to be is that hopefully we get really great information that will help us improve the quality of care for patients. That is our overarching goal.

How has the reaction been amongst the clinics?

Lena: Great, they're all excited to do it. It's just resources; it takes time to implement something like this. Each clinic has to take time out of their busy schedules to actually put it in, submit an ethics application. But that's why I'm here: to help guide

them and present them with the general protocol—which they obviously adapt to their own clinic before submitting to ethics—and I help them with their basic IT hurdles.

Lisa: Our goal is to get two to three clinics per year on board. We've been meeting that target consistently so far, and it looks like we will continue to do so up until 2019. ■

“But ultimately, really, what the big change is going to be is that hopefully we get really great information that will help us improve the quality of care for patients. That is our overarching goal.”



Research Corner:

FOX_{M1}: A Potential New Therapeutic Target for Pulmonary Hypertension



Alice Bourgeois became the first recipient of the PHA Canada *Mohammed Family PH Research Scholarship* in 2017 for her studies in pulmonary arterial hypertension (PAH), under the supervision of Dr. Olivier Boucherat, PhD. Her project is associated with Laval University, Department of Medicine, IUCPQ Research Centre, where she is currently completing her Master's in Molecular Medicine. She has always had a strong interest in understanding the fundamental mechanisms that cause disease, which is why she chose to pursue her graduate studies in PAH. After two years of working as a summer student for the PAH Research Group at Quebec City's Cardiology and Respiriology Institute's Research Centre, she remained involved with the PAH Research Group for her graduate studies. Her work is focused on understanding the scientific mechanisms leading to the development of PAH and investigating new potential therapeutic options.

Pulmonary hypertension (PH) is defined by an increase in the blood pressure in the pulmonary arteries (blood vessels that feed the lungs), because these arteries are narrowed and sometimes completely closed. Narrowing of the pulmonary arteries is the result of different things, but two of the major causes are spasm and the build up of cells and scar tissue (known as vascular remodelling). Current available treatments for PH generally do not completely relieve symptoms or improve the pulmonary artery narrowing in the long term. Most medical therapies currently in use are vasodilators that relax pulmonary arteries, targeting one of the components responsible for narrowing the arteries. However, it is not clear if current PH medications are effective in reversing vascular remodelling. Thus, there is a need to better understand pulmonary artery remodeling in PAH and to identify new therapeutic targets and develop new treatments in order to be able to improve the life expectancy of patients. This was the starting point for my research.

It is known that the wall of the pulmonary arteries is partly made of muscle, specifically smooth muscle cells. In the pulmonary arteries of a patient with PH, these smooth muscle cells are under much stress in their environment, as they face conditions that are unfavourable to cell survival. Surprisingly, these smooth muscle cells do survive and even reproduce despite this environmental stress. These are characteristics that are similar to cancer cells. Although PH is clearly not a cancer, the similarities open up the possibility of better understanding PH and perhaps using cancer treatments in PH patients.

In my project, we were interested in FOX_{M1}, a chemical found inside cells that is known as a transcription factor. Chemicals like FOX_{M1} control cell survival, growth, and function by turning on genetic information in the DNA. In cancer

cells, there are higher levels of FOX_{M1} compared to healthy cells and this higher level causes cancer cells to reproduce uncontrollably rather than undergo normal death through a process known as apoptosis. Since FOX_{M1} is an essential transcription factor for normal development and growth of the lungs, we hypothesized that higher levels of FOX_{M1} in PH could cause the abnormal survival and growth of smooth muscle cells in the walls of pulmonary arteries.

To find this out, we obtained the diseased lungs from PAH patients after they had undergone lung transplantation and isolated the pulmonary arteries. We found more smooth muscle cells in these PAH pulmonary arteries and—when we treated the smooth muscle cells with a drug that blocks the action of FOX_{M1} (thiostrepton)—we found that there was a significant decrease in cell reproduction and an increase in normal cell death. This suggested that thiostrepton could reduce pulmonary artery vascular remodelling and scarring in PAH. Next, we tested the therapeutic potential of thiostrepton in preclinical animal models of PH. Studies in two of these models allowed us to observe similar results: thiostrepton decreased reproduction of smooth muscle cells in pulmonary arteries and increased the normal death of these cells, resulting in a decrease in vascular remodeling and less narrowing of pulmonary arteries. Very importantly, there was also a decrease in the severity of the PH. Although further studies are needed, we believe that FOX_{M1} may be a potential new future treatment option for patients with pulmonary hypertension. ■

Contributed by: Alice Bourgeois, PH Research Scholarship Recipient, Laval, QC



This November, show your

Life In Purple

#PHAwareness Month 2018

Celebrated annually throughout the month of November, PH Awareness Month is an international event that raises awareness of pulmonary hypertension (PH). Every November, people living with PH worldwide, along with their loved ones and care providers, unite to raise awareness of the disease and the need for earlier diagnosis, more effective and accessible treatments, and—ultimately—a cure.

NOW IS A GREAT TIME TO START
THINKING ABOUT NOVEMBER!



1. Organize an event to help raise awareness in your area!
2. Request that your local government declare November as PH Awareness Month!
3. Join the campaign on social media using #LifeInPurple
4. Wear purple for a week, then challenge your friends and family to do the same!
5. Think about making a donation to the Canadian PH community!



Get involved by visiting
phacanada.ca/LifeInPurple2018

FREE for PH patients
& primary caregivers!

CONNECTIONS

Subscribe to the Canadian PH Community's Official Magazine!

Published twice a year, *Connections* is a bilingual publication that brings together the Canadian pulmonary hypertension community to inform, support, and celebrate one another.

Subscribe

- **Lifetime subscription** — FREE for PH patients & primary caregivers (limit one per household)
- **1-year subscription (two issues)** — \$25.00
- **2-year subscription (four issues)** — \$40.00

To learn more & subscribe online, visit www.phacanada.ca/connections.

Contribute

Connections is your publication. Tell us about your support group or recent event; share your story or tell us about a phenomenal caregiver in your life; or let us know how you cope with PH on a daily basis. If you're not comfortable writing your story, contact us, we'll interview you and write the story for you. Let your voice be heard, that's what *Connections* magazine is all about!

Deadline for submissions for the next issue: January 15, 2019.

Content Disclaimer

The content featured in *Connections* magazine is created by members of our community, and the information is checked for accuracy to the best of our ability. However, each person's PH story is unique, so what works for one individual may not work for everyone. If any information in *Connections* doesn't seem correct to you, please let us know so that we can verify it. Most importantly, always check with your PH team before making any lifestyle or treatment changes.

**ALMOST 1 IN 3
PEOPLE DIE FROM
CARDIOVASCULAR
DISEASE** | **OUR
SCIENTISTS
WON'T
ACCEPT THAT**



According to the World Health Organization (WHO), cardiovascular diseases are the number one cause of death throughout the world. That's why we're investigating treatments to manage heart failure, as well as prevent myocardial infarctions and strokes. Which heart patient wouldn't love to benefit from that?

To find out how our innovations are helping to change lives for the better, visit www.bayer.ca

