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

CONNECTIONS

The Official Magazine of the Canadian PH Community

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For subscriptions, contributions, or inquiries, please contact us at connections@phacanada.ca.

Editor in Chief
Mariane Bourcheix-Laporte, Communications Associate

Contributing Editor
Jamie Myrah, Executive Director

Translation
Denise Bérubé
Rahul Bhundhoo
Mariane Bourcheix-Laporte
Jean Dusseault
Elizabeth Lim

Art & Design
Mariane Bourcheix-Laporte
Olga Roberts

Printing
Blanchette Press

Contributing Writers
Mariane Bourcheix-Laporte
Jennifer Gendron
Tina Giroux-Proulx
Dr. Nathan Hambly
Arnold Hull
Carson King
Adam Kingz
Jamie Kretzschmar
Serena Lawrence
Gail Nicholson, RN
Roberta Massender
Dr. Sanjay Mehta
Jamie Myrah
Valérie Plouffe
Dr. Charles D. Poirier
Sylvia Rinaldi
Mohamad Taha
Virginie F. Tanguay
Jeannie Tom

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How Do We Choose the Optimal Treatment for Each PAH Patient?

Hello PHriends,

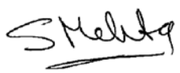
In a serious, progressive, and often fatal illness such as pulmonary arterial hypertension (PAH), each patient living with PAH, their healthcare providers, and caregivers want to find the optimal treatment, i.e. the treatment that, all things considered, is right for that individual. PAH treatments offer various benefits: fewer and less severe symptoms; a better ability to be active and function in everyday life; less frequent complications such as admission to hospital; a better health-related quality of life (HRQoL); and hopefully longer survival. As for any medical treatment, there are side-effects and risks associated with all PAH treatments, ranging from minor and troublesome (eg. mild nausea, occasional headache) to more serious, such as an infection of the blood acquired through the indwelling permanent catheter (tubing) inserted into a neck vein, which is required for continuous infusion of intravenous medications.

Based on consideration of these risks and benefits, the patient and their PH physician come to a mutual decision as to the best choice for initial therapy for that patient, as well as future treatment options. Although each PAH patient wants to improve, they don't make the same choices. For example, less than 200 Canadians are currently receiving one of the available complex intravenous or subcutaneous infusion therapies, despite evidence that these are the most effective treatment options for PAH. In contrast, thousands of Canadian PAH patients are currently receiving effective oral medications, either alone or in various combinations of two or three drugs.

It is a basic tenet of medicine that each patient is an individual with different goals and values in life, health priorities, and needs. One patient may value longer survival over all else and choose the most aggressive treatments, accepting more side-effects. Other patients may value minimal side-effects and opt for more conservative, simpler treatment options with limited clinical benefits, accepting greater risks of illness progression and shorter survival. As such, a PAH patient is clearly the only person who can decide the optimal treatment for their illness.

It is our responsibility, as healthcare providers, to help each patient and their caregivers understand the nature and potential benefits of available treatments as well as the possible side-effects and risks. We work hard to help each of our patients improve as much as possible in the hope that they will be as well as they can be for the longest time. However, we also want each patient to do what is best for them, and so we must and do accept that some PAH patients don't always opt for what we may consider as the "best" treatment options. We accept this paradox because the decision is not our; the illness, life, and choices are the patient's.

There is increasing focus in the PH medical community on understanding each patient's experience of their PAH, choice of optimal therapy for their illness, and assessment of their HRQoL. It is important that healthcare providers understand a patient's ideas, values, and choices in order for them to provide the best care. Moreover, clinical research studies in PAH are increasingly considering the patient perspective through measuring patient-reported outcomes (PROs), including self-reported symptoms and benefits. Similarly, in PHA Canada's ongoing advocacy work encouraging government agencies to provide access to all Health Canada-approved PAH therapies to all PAH patients, the voice of each individual patient and caregiver sharing their experience living with PH is vital to our success. We invite you to share your stories, goals, values, priorities, and needs with your healthcare providers and the broader PH community. This will help you deal with your own illness as well as benefit many others in your community. If you don't know how to tell your story, we encourage you to find your voice by learning from the inspiring stories of others.



Sanjay Mehta, MD, FRCPC, FCCP

Director, Southwest Ontario PH Clinic, LHSC – Victoria Hospital, London

Chair, PHA Canada Board of Directors

Message from the Executive Director: Empowering the PH Community

Each of us takes on the role of "patient" during our lifetimes. For some, it will be a small part, perhaps reoccurring every so often, but never dominating the spotlight for too long. For others, it will become a permanent state of being, one that must be integrated alongside the other big roles in life: partner, parent, sibling, child, etc. At the same time, life also frequently demands that we be caregivers, often compelling us to occupy both patient and caregiver roles simultaneously. As both patient and caregiver, we are called upon to learn complex new information, make complicated life-altering decisions, and support people we love deeply through pain, sorrow, and fear. It can leave us feeling overwhelmed, inadequate, and powerless to act in the interests of our loved ones, or even ourselves.

Health-care providers, support workers, and community advocates often point to the concept of "empowerment" as a key way to counter such feelings and equip patients and caregivers with the tools they need to address these challenges. At PHA Canada, our stated mission is to ***empower*** the PH community through support, education, advocacy, awareness, and research. So, what do we mean by ***empower the PH community***? What power is it that we want patients and caregivers to be able to wield? And why is it so important?

The answer is both plainly simple and deceptively complicated. On the surface, empowerment is about people taking control over their own lives, acting on their own authority—either individually or as a group—to achieve their own goals. And yet, our unique capacity for such self-determination is constantly threatened and constrained. How do we know the information we seek is appropriate and reliable? How do we decide when we don't have the same choices as someone else? How do we take action when we feel so exhausted?

This is why PHA Canada exists: to help you live your best possible life with pulmonary hypertension, according to your own goals and values. We provide educational materials to newly diagnosed patients so they can learn what's happening in their body and make more informed decisions about their care. We share information about some of the conditions that are associated with PH (insert pages X-XX) so PHighters can better understand how the disease affects everyone differently. We invite the community to participate in regional symposiums (insert page X-XX) so you can share your experiences and build supportive relationships with one another. These resources are here to help you gain the confidence and abilities you need to successfully manage the effects of living with PH.

Not everything we do to empower the PH community is as obvious as our printed resources and community events. Much of our time is spent behind the scenes, working with others to create an environment where patients and caregivers can leverage their abilities and successfully influence the direction of their own lives. For instance, when advocating in partnership with Scleroderma Canada for PH patients to have equitable access to all Health Canada-approved treatments, some of our work is focused on creating ways for people to be able to advocate on their own behalf, such as with the email and advocacy tools available at www.TakeActionPAH.ca. However, much time and effort is also spent meeting directly with politicians and policy makers to ensure your voices are being heard and considered in the decision-making process. In facilitating this direct exchange between the PH community and decision makers, we help educate both groups so they can communicate more effectively and find common ground.

Of course PHA Canada does none of this work by itself. You—the PH community—are what drives us, from the leadership of our Board of Directors to the guidance and support of our specialty advisory committees and community volunteers across the country. Without you, in 2016 we wouldn't have had our biggest November PH Awareness Month yet (pages 6-9), awarded two new PHA Canada Paroian Family PH Research Scholarships, or developed new resources on CTEPH, keeping kids with PH active, and early diagnosis. We look forward to working with you throughout 2017, whether you choose to send a letter to your provincial representatives about access to new treatments, share your PH journey with your local media, volunteer for an event or committee, or become a donor so that we can continue to deliver on our mission. As Dr. Mehta said, "we invite you to share your stories, goals, values, priorities, and needs" because it is only by working together on what matters most to you that PHA Canada can fulfill its mission of empowering the PH community.

Jamie Myrah

Executive Director, PHA Canada

PH Awareness Month: Periwinkle Power Demonstration

Every November, PH patients, caregivers, supporters, and health care professionals across the country and around the world take part in awareness-raising activities that put a face to PH, make the signs and symptoms of the disease known, and bring attention to how PH impacts individuals and families. Last November, our community put on a PHabulous demonstration of periwinkle power with awareness events and fundraisers organized throughout the country. We thank all the event organizers, volunteers, sponsors, and donors for supporting the cause and helping us create a better life for all Canadians affected by PH. Here is an overview of PH Awareness Month 2016 events.

THIRD ANNUAL RUN/WALK, SWIM OR BIKE/SPIN PH CHALLENGE (October 24th to November 3rd, Bradford, ON)

For the third year in a row, athletes in Bradford put on their walking shoes and swimming goggles in support of PHA Canada. Organized by PHA Canada Board Member Ruth Dolan, this weeklong fundraiser challenged participants to run, walk, swim, or bike/spin as many kilometers as possible to collect funds in support of our activities. With over $2,700 raised, it's safe to say that participants took on the 2016 PH Challenge head on!

Thank you Ruth for your unwavering commitment to the PH community.

From left to right: Stephanie Uren and Bradford Mayor Rob Keffer; PH Challenge participants; Councillor Gary Baynes, Ruth Dolan, and Mayor Rob Keffer; and Zarco Tesic and Carol Comeau, who were awarded 1st place for swimming.

FOURTH ANNUAL DINE FOR THE CAUSE AND SILENT AUCTION (November 2nd, Ottawa, ON)

In Ottawa, raising PH awareness comes with fine dining! PHA Canada power couple Teri and Harry Kingston organized a joint charity dinner and silent auction with the Ontario Lung Association at Biagio's Italian Kitchen. For the fourth consecutive year, supporters in the Ottawa region came together for a special evening focused on lung health awareness and community support. We raise our glass to the Kingstons and Ottawa supporters who raised $700 to the benefit of PHA Canada's programs and services. Congratulations on organizing another successful event!

Supporters enjoying an evening of fine dining and lung health awareness.

BE BRAVE AND SPARKLE (November 4th, Newmarket, ON)

The Berdan family is new to the PH community (read their story pages 10-11), bringing loads of passion for raising awareness and finding a cure! With the support of their extended PHamily, the Berdans held a sparkling PHundraiser on November 4th, raising awareness of PH and celebrating their daughter Mia's strength. Fun games, dancing, face painting, and a generous dose of sparkles made this event one to remember! We thank the Berdan-Clan for their incredible energy and commitment to the cause. With over $5,600 raised through this event, you have made the whole PH community sparkle!

Activities for the whole PHamily were organized at the Funnery Play Park & Café, the Move Fitness and Dance Studio.

SAIL-O-THON BENEFITTING PHA CANADA (November 4th, Vancouver, BC)

Our Communications Associate, Mariane Bourcheix-Laporte, is passionate about the work she does every day in support of the PH community. She is also an avid sailor and decided to combine both passions by challenging herself with a sail-o-thon benefitting PHA Canada. Strong winds, big waves, and cold weather made her 40-Km sail a real challenge and a truly impressive achievement! Thanks to the support of friends, family, and the PH community, Mariane's sail-o-thon raised over $2,600 in support of PHA Canada's activities. Nothing takes the wind out of her sails!

It was chilly out on the water for Mariane's Sail-o-Thon!

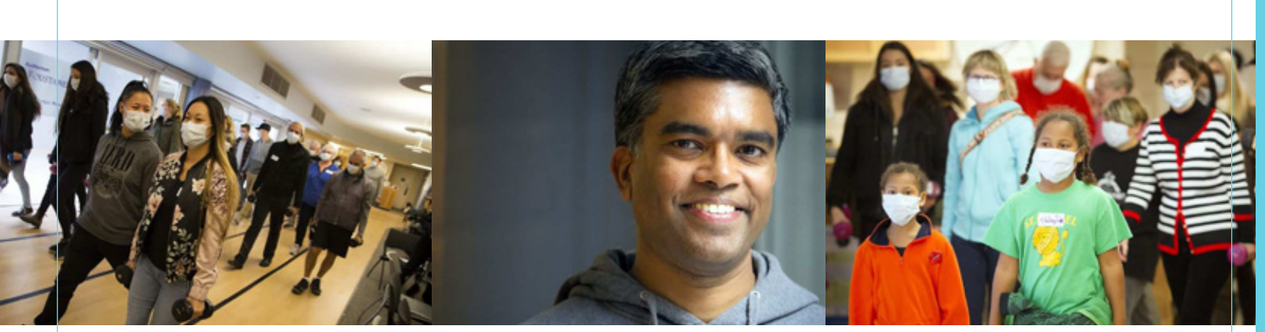
FOURTH ANNUAL MASQUERADE BALL (November 5th, Calgary, AB)

PH Nurse Gail Nicholson has made it her mission to raise awareness for the "real people behind rare disease" by organizing the Annual Masquerade Ball fundraiser in support of the PH community. A dazzling event complete with costumes, masks, and musical and dance performances, this fundraiser turned invisibility on its head by celebrating the people who fight against an invisible illness. Thank you Gail for your continued commitment to the PH community.

Supporters in Calgary had a dazzling time at the Masquerade Ball!

6-MINUTE WALK FOR BREATH (November 12th, Ottawa, ON)

The Ottawa PH clinic held its annual ***6-Minute Walk for Breath*** fundraiser and community gathering at the University of Ottawa Heart Institute. PH patients, caregivers, and supporters participated by walking the Institute's 6-minute walk test route; those without PH wore masks and used weights to get a better sense of what doing the test is like for people with PH. Thank you to PHAC Board Member and Nurse Carolyn Doyle-Cox for organizing the event and for inspiring PHighters in the Ottawa region to raise over $3,600 in support of PHA Canada's programs and services.

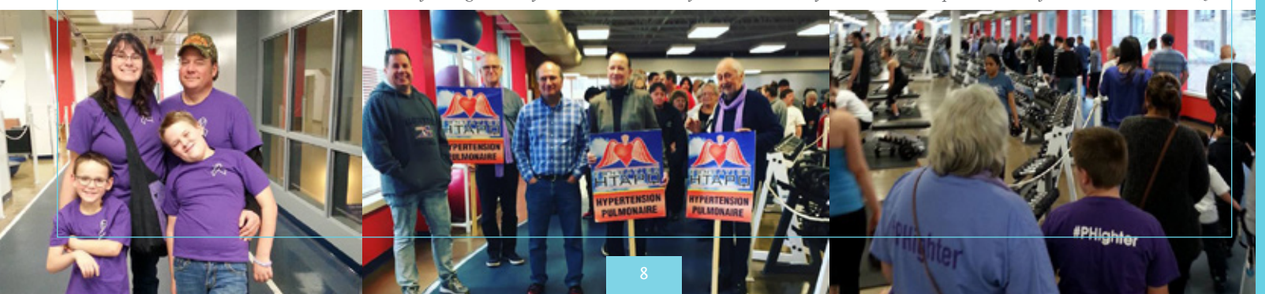


Photos of the Ottawa 6-Minute walk for Breath sourced from "DND contractor needs to keep working to pay for life-saving drugs," Ottawa Citizen, November 12, 2016.

FIFTH ANNUAL MONTREAL PH AWARENESS WALK (November 27th, Montreal, QC)

Members of the Quebec PH community came together at the YMCA in downtown Montreal to participate in this community gathering and fundraiser benefitting the Montreal Jewish Hospital. Our Executive Director, Jamie Myrah, was in town for the event and walked alongside Fondation HTAPQ members and Montreal PH clinic professionals. Thank you to Nurse Lyda Lesenko for organizing the event and being a positive force in our community.

From left to right: Wendy Bedard, Kendall Tracy, and their sons; Lyda Lesenko and representatives of the Fondation HTAPQ.



OTHER EVENTS

Awareness tables, community gatherings, and municipal proclamations are some of the other ways our community helped spread PH awareness last November. PHighters across the country also participated in our Lend a Hand campaign to educate those around them about PH. Participants asked friends and family to contribute tokens of support by signing or tracing their hands on specially designed posters and then making a donation to PHA Canada. Thank you to everyone who lent us a hand by raising awareness of PH and funds to support our programs!

PHOTO CAPTIONS

PH AWARENESS PSAs

In 2016, PHA Canada initiated its most significant public education campaign yet with the broadcast of two new bilingual video PSAs on local and national television! Thanks to our ongoing collaboration with filmmaker Tyler Gamsby, we produced 30-second video PSAs that aim to raise public awareness of the symptoms of PH and the impacts of the disease on patients and families. We received incredible support from the following stations who aired our video PSAs throughout the month: Global BC, Rogers TV Ontario, Rogers TV New Brunswick, Shaw TV Calgary, Shaw TV Vancouver, Shaw Multicultural Channel, and MaTV Quebec. Coastal Radio (Glace Bay, NS) and Roundhouse Radio (Vancouver, BC) also aired our audio PSA throughout the month of November.

Stills from our two video PSAs. Far left: Justin Price and Natalie Roy.

Supporters in Newfoundland raising awareness at Grand Falls-Windsor City Hall for PH Awareness Month.

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On Being Brave and Sparkling: The Berdan Family's Story

In the summer of 2016, the Berdan Family was shaken by the news of daughter Mia's PH diagnosis. Since then, they have been active on multiple fronts of the PHight against PH; they have become PH awareness champions, have raised funds to support PHA Canada, and have learned to draw from their PHamily's strength to face challenges and care for Mia.

Just before Mia turned 10, we started to notice that she wasn't breathing normally. She had never been a sporty kind of kid, but we started to worry when she stopped being able to dance, sing, or even walk a short distance without stopping to catch her breath. She was diagnosed with asthma and prescribed puffers. However, 2 months later, Mia started waking up in the middle of the night with chest pains, feeling as though she was having a heart attack. She began fainting and seizing, had no energy, her skin looked greyish, and her lips were slightly blue.

In the course of a few months, she went from belting out her lines in a musical theatre competition to barely being able to walk across the stage. We couldn't get on board with the doctor's asthma diagnosis so we sought a second opinion from the Newmarket South Lake Hospital, where thankfully a doctor knew of PH and referred us to a cardiologist. We were referred to the pediatric PH team at the Toronto Sick Kids Hospital and, after what felt like a trillion tests, Mia' PH was confirmed. Hearing a doctor tell you that your child has a progressive lung disease that is affecting her heart, and then hearing them say there is no cure for it is a parent's worst nightmare.

PH has impacted our family's life in many ways. First, we had to understand that with PH there is no normal; normal literally doesn't exist in our world. "Live every day as though it was your last" has become our family's new motto, although it's easier said then done. We are starting to accept that we really can't make plans, because they can get cancelled in an instant,

This photo: Mia's diagnosis has made the Berdans closer than ever.

Below: The Berdan-Clan celebrating periwinkle power and raising awareness of PH at the Newmarket Santa Claus Parade last November.

so we've become "last minute joiners." We are learning as a family what is important and what isn't and choose outings and activities based on Mia's fragile health. We now focus on relationships and the people we love. As a result, we spend more time together and talk, laugh, smile, and hug more. As a family we communicate much better and it seems that there is hardly any yelling in our house anymore. Everybody helps everybody else; we do things together and everyone has a voice.

We became involved with PHA Canada soon after Mia's diagnosis. As soon as my husband heard the words "pulmonary hypertension," he wanted to learn everything he could about the disease, did some research, and came across PHA Canada. Being a former Activity Director, organizing social activities is my second nature and I wanted to get involved in raising awareness of PH by getting the word out there. I believe that the more people know about PH, the less patients will be given the wrong diagnosis and continue to suffer; the more people know about PH, the more money they can donate towards awareness, research, and finding a cure.

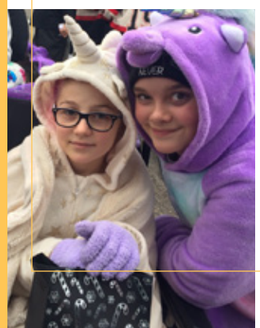
So last fall, with the help of friends and family, the Berdan-Clan organized a number of awareness and fundraising activities. We gave away information along with candy at Halloween; we organized the Be Brave and Sparkle PHundraiser for PH Awareness Month; marched to raise awareness at our local Santa Claus parade; held an Awareness Assembly; organized a benefit Christmas Concert; and turned teacher holiday gifts into an occasion to raise funds. The last few months of 2016 were a very busy time for our PHamily! None of it could have happened without the help of our PHabulous PHriends, whom we know love our family so much they would do whatever it takes to drop kick PH into extinction!

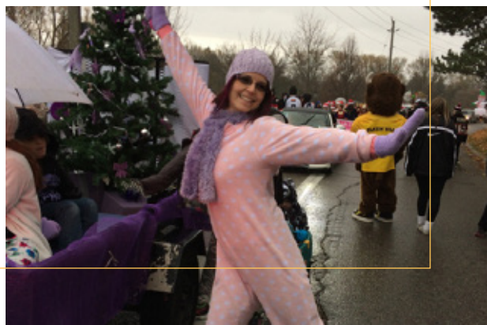
Organizing these events, I was overwhelmed by people's generosity and I am still surprised by the support we continue to receive. Having a kid with PH can be pretty isolating, especially during the winter months with sickness everywhere and germs spreading like wildfire, yet we never really feel alone. People seem to have rallied around us in our time of need; I call them my "feel-better-fairies." They've supported us in every way imaginable and have made it possible for us to care for Mia more easily.

My hope for the not-so-distant future is that we find a cure for PH and I believe it's going to take a huge village to win this PHight. In the mean time, my hope for Mia is the same as my hope for her little sister, Allie. I hope she gets the chance to become everything that she was meant to be, that she'll always be comfortable with exactly who she is, and that she'll love herself enough to confidently make good life-choices.

My family has learned that blood doesn't make people PHamily, unconditional love, support, and a common goal do. We have a huge PHamily!

Contributed by: Chris Berdan, PH caregiver, Newmarket, ON





Susan and Jim Bailey are facing the challenges of life with PH together.

How Not to Take It Sitting Down

Susan Bailey was diagnosed with PH just over a year ago, but she has already shown that she is a fierce PH advocate, raising awareness everywhere she goes and becoming a star fundraiser last November for the 2016 Ottawa 6-Minute Walk for Breath.

I was diagnosed in late February 2016 when my symptoms deep cough, edema, fatigue, fainting, and shortness of breath became unbearable. A savvy ER doctor initially diagnosed my PH after performing a number of tests, including an ECG, an echocardiogram, and blood work. He immediately referred me to the Ottawa Heart Institute's PH clinic, where I am now treated.

LIVING WITH PH

Since my diagnosis, my husband Jim's life and my life have changed. PH has seriously curtailed our travelling. At one time, we were fortunate enough to go on cruises, travel to Europe, or head south for a few weeks every winter. My mobility is now limited; I am no longer able to shop for hours, do groceries, or run errands. Socializing is largely contingent on how well I am feeling on any given day or how low my oxygen levels (SATS) are. However, out of all the situations I have had to adapt to in the last months, three things stand out as exceedingly difficult: completing extensive paperwork to receive funding for very expensive drugs, depending on my husband and friends for most activities, and transporting oxygen everywhere I go to ensure that I am not without it for long.

I get through the challenges associated with my "new normal" by being positive, staying realistic, and relying on my strong faith. Accepting my situation while remaining very hopeful for a positive future and reaching out to others also play huge roles in ensuring my well-being. I regularly attend Ottawa PH support group meetings, go to counseling, and know I can count on support from my church community and friends. Moreover, I am lucky to have the support of my "earth angel" my dedicated husband of almost 37 years, who is standing by my side throughout this challenge.

BEING A PH ADVOCATE

I am not one to take anything sitting down, so after my diagnosis I said: "if I must have this disease, then put me to work!" Raising awareness of PH has now become my job; I educate those around me wherever I can. Facebook is a great resource for getting the message out! During PH Awareness Month, I was also able to share my story in the media with television and radio interviews. I will continue to advocate as long as I have the strength to do so.

Participating in the Ottawa 6-Minute Walk for Breath at the University of Ottawa Heart Institute was a great way to raise awareness of PH as well as funds to support PHA Canada. After signing up for the event, I reached out to my network and asked for support. I am a retired teacher and have a large network of former colleagues and students as well as many caring friends. I was surprised by their generosity, although I should not have been. I set myself a very modest fundraising goal to begin and quickly had to change it. In the process, I was overwhelmed by the kindness I received.

STANDING UP TO LOOK AT THE PHUTURE

I refuse to give up on Jim and I's dream to go back to France and spend at least a month in the country that we consider to be our "heart home." The many advances that have already been made in life-sustaining PH therapies also make me hopeful. My hope for those of us afflicted by this vile condition is that an outright cure be found... soon!

Contributed by Susan Bailey, PH patient, Ottawa, ON

Monter la montagne ensemble

Marie Andrée Malette is a fierce PH advocate

Ma mère, [nom] a reçu son diagnostic d'hypertension pulmonaire en 2010, après avoir souffert de difficultés respiratoires pendant près d'un an suite à une infection pulmonaire reliée au virus H1N1. C'est à la clinique d'HTP de l'Institut de cardiologie de l'Université d'Ottawa qu'elle a reçu son diagnostic et a été traitée par la suite. Après cinq ans de lutte, elle est décédée subitement le 19 novembre 2015.

L'impact de la maladie sur la vie de ma mère a été immense : elle a dû cesser les activités qu'elle aimait tant et arrêter de travailler. L'HTP a grandement éprouvé son estime de soi et son indépendance financière ; en plus de ses symptômes physiques, elle a souffert d'anxiété et de dépression liées à la maladie. Notre famille l'a accompagnée au long de cette épreuve. Mon frère et moi, ainsi que d'autres membres de la famille, ont été ses aidants naturels. Étant infirmière, je me suis impliquée de près avec son équipe médicale, communiquant régulièrement avec son personnel soignant.

Lorsque ma mère a reçu son diagnostic, je lui ai dit : « ça sera comme une grosse montagne qu'on montera ensemble ». Lorsqu'elle est décédée, j'ai voulu monter la montagne pour elle et remercier la clinique d'HTP d'Ottawa pour les merveilleux soins qui lui ont été donnés. J'ai donc entrepris de grimper le Mont Marcy dans les Adirondacks cet été au profit de l'Institut de cardiologie de l'Université d'Ottawa, qui abrite la clinique où ma mère a été traitée.

Ce défi a plusieurs objectifs : m'aider avec mon deuil, remercier la clinique d'HTP d'Ottawa, faire avancer la recherche et les traitements, et sensibiliser à l'importance des interventions précoces et parfois agressives pour atténuer la progression de la maladie. Ce défi me permet de récolter des fonds pour soutenir l'Institut de cardiologie de l'Université d'Ottawa, mais c'est aussi un moyen pour moi de faire de la sensibilisation auprès du public. Je souhaite faire valoir l'importance du soutien à domicile afin que les personnes qui vivent avec des maladies chroniques comme l'HTP puissent conserver leur autonomie ; l'importance de certains traitements qui peuvent acheter du temps ; l'importance du don d'organe ; l'importance d'écouter les besoins des patients ; l'importance de l'aide financière aux gens qui sont atteints de maladies chroniques sévères ; et l'importance d'être à l'écoute de leurs besoins en santé mentale. Ce sont aussi des messages que je désire transmettre aux gouvernements.

Heureusement, j'ai espoir en l'avenir! C'est pourquoi je m'implique au près de la cause. Ensemble nous pouvons faire changer les choses !

Contribution de Marie Andrée Malette, aidante et militante, Gatineau, QC

From left to right: Marie Andrée's mother, name photographer with her son; Still from Marie Andrée's CTV News Ottawa interview, December 19, 2016.

Overview of the 2016 Alberta PH Forum

On Saturday, October 01, 2016, over 50 PHighters gathered in Red Deer for PHA Canada's first Alberta PH Forum! Patients and their caregivers, along with medical professionals from Edmonton, Calgary, and Red Deer, came together to expand their knowledge about living with PH, and connect with PHriends both old and new.

The day started with a session on Support Systems for Coping with Chronic Illness, presented by Registered Social Worker Kirsten Lambert. Kirsten, who has worked with families affected by PH at the Peter Lougheed Centre in Calgary, spoke about the different kinds of supports available, from professional supports to personal and self supports. Special attention was given to the important role played by peers and support groups when dealing with a chronic condition such as PH. Next up was a skills building workshop on Eating Well with Pulmonary Hypertension by Lauren Reiger, Registered Dietician from the Heart Function Clinic at the Red Deer Regional Hospital. Thanks for helping us better understand those nutrition labels, Lauren!

After a chance to get to know one another better over a low sodium lunch, the participants split up into two breakout sessions. The families of kids with PH attended an intimate panel discussion on Pediatric Care with Dr. Ian Adatia from Stollery Children's Hospital, Karen Janz, Remodulin Nurse with McKesson's Patient Support Program, and PH-parent Shannon Reitor, accompanied by her little PHighter Adam. Meanwhile, the panel discussion on Adult Care featured: PH Clinical Fellow Dr. Vikrum Gurtu; Dietician Lauren Reiger; and PH Nurses Gail Nicholson, Josette Salgado, and Cheryl Salvador.

The day wrapped up with two more large group presentations. The first and most popular of the day was a fascinating look into The Future of PH Therapies with Dr. Gurtu. Lastly, a panel of patients and caregivers shared their Tips & Tricks for Living Well with PH. Special thanks to Lynn-Marie Cox, Ted Earl (and his widow Pam), Marcin Godzik, Stephanie Ricci, and Lynn Williams for sharing their wisdom with us.

PHA Canada gratefully acknowledges the support and contributions of the many people who have made this event possible, especially our participants, who came from throughout Alberta and even as far away as BC! Thank you to Alberta's pulmonary hypertension clinics, as well as our wonderful volunteer steering committee members: Lynn-Marie Cox, Marcin Gozdzik, Gail Nicholson, Stephanie Ricci, Cheryl Salvador, and Heather Zloty. Thank you also to our event volunteers Katherine, Paige, and Preston Nicholson and to all the speakers for sharing their time and expertise. Finally, we also thank Unither Biotech for their 2016 Silver Level Sponsorship of our support and education programs, and in particular for this first ever Alberta PH Forum.

Next up: Ontario & the Eastern Region!

Contributed by: Jamie Myrah, Executive Director, PHA Canada

Above: Participants learned to manage sodium and fluid intake.

This photo: Forum presenters with PHA Canada Executive Director, Jamie Myrah. From Left to right: Cheryl Salvador, RN; Josette Salgado, RN; Gail Nicholson, RN; Dr. Vikram Gurtu; Kristen Lambert, and Registered Social Worker.

"HTAPQ SYMPOSIUM AND CHEESE SALE" PLACE HOLDER

PHA Canada Eastern Regional Symposium

Save the date: Toronto | Oct. 13-14, 2017

Mark your calendars for our first Eastern Regional PH Symposium, which will take place ***October 13-14, 2017*** at the Westin Prince Hotel in Toronto. Join us for Friday night for a special welcome reception and Saturday for a full day of educational sessions featuring medical experts and your PH peers. We are happy to announce that PHighters living in, or east of Manitoba will be eligible to apply for travel scholarshipsstay tuned for more details. More information on this event will be announced in the coming months, including further details on scholarships, registration, volunteer opportunities, and program updates. Stay up-to-date by visiting www.phacanada.ca/easternsymposium2017regularly.

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 will celebrate its tenth anniversary. In preparation for 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.

"To me, PHA Canada means education and support. *When I was first diagnosed and trying to cope with the implications of PH, PHA Canada was there for me with clear explanations and resources. Later, it is through this great organization that I learned how to advocate on behalf of all PH patients for access to treatment. I am grateful to know that PHA Canada is there for patients like me, and I thank them for all of the work they are doing on behalf of Canadian PHighters."*

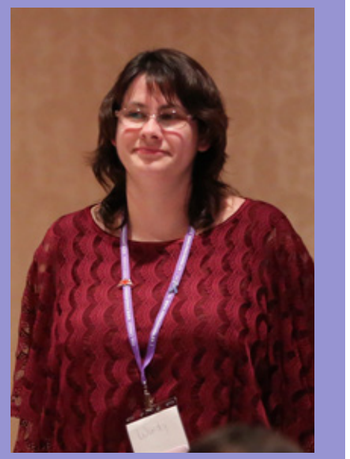
—Beth Slaunwhite, patient, Halifax, NS

"It means community and knowing that there are others that 'get' me because they experience similar things. *Together we have a louder voice."*

—Juergen Buettemeyer, patient, Ingersol, ON

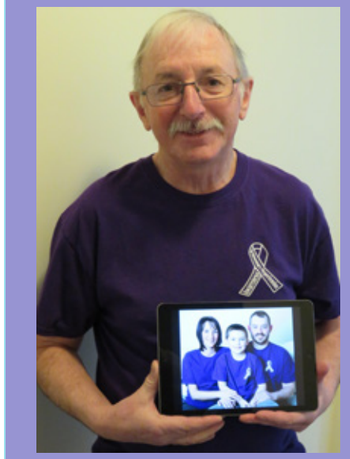
"It means hope, love, acceptance, and PHamily. *We are extremely grateful to PHA Canada for making our lives better!"*

—Wendy Bedard, caregiver, Mansonville, QC



"To me, PHA Canada means that there are people who care *and are fighting for me to survive. PHAC has impacted my life by letting me know I am not alone in this. The most valuable service PHA Canada has offered me is support. I know I can call or email at anytime if I need something and they will try their best to help me with whatever I am looking for."*

—Jas James, patient, Cobble Hill, BC



"In the initial stages of our PH journey, we had an immediate need for education *about the disease and connection with people facing the same PHight. PHA Canada provided the medium to connect with others living with PH and discover what it meant to them."*

—Carson King, father-in-law of lung transplant recipient Wendy King, Grand Falls-Windsor, NL

"To me, PHA Canada means community. *Despite so few people in Canada having been diagnosed with PH, such a strong and dedicated community of patients, caregivers, and professionals has been established. It is comforting to be connected with others who truly understand how PH can affect several aspects of life.*

The most valuable service that PHA Canada offers is education and awareness. Before my mom was diagnosed, we had never heard of PH. We value the educational pamphlets and flyers that PHA Canada creates and sends us to distribute at the fundraisers we organize. In order to find a cure for PH, more people outside of the PH community need to know about the disease. That awareness stems from the educational resources PHA Canada creates."

—Renae Mohammed, caregiver, advocate, and fundraiser, Ajax, ON

"To my family, PHA Canada means hope... *We are not alone in our fight against PH"*

—Sarah Platnar, mother of PHighter Isabelle, PHA Canada Pediatrics Committee Chair, Pickering, ON

Knowing Where to Look: Conditions Associated with PH

Currently in Canada, it takes more than two years for many patients to get diagnosed with PH. This statistic is unacceptable, especially given that without treatment the average life expectancy of someone with PH is less than three years. Because PH is a progressive disease, the earlier patients are treated, the better their chances of managing their symptoms and maintaining their quality of life. Unfortunately, lack of awareness of PH can cause significant diagnosis delays, with the result that 75% of patients have advanced PH by the time they are diagnosed.

One of PHA Canada's strategic priorities is improving the statistics mentioned above. We believe that early diagnosis of PH is critical to optimal treatment, which is why we have developed a number of awareness initiatives through our Early Diagnosis program. With public service announcements broadcast on television and the radio, we are encouraging Canadians experiencing symptoms of PH to talk to their doctor about the disease. We are also reaching out to the medical community with new resources designed to help physicians identify the signs and symptoms of PH, order investigative tests if they suspect PH, and refer patients to a specialized centre to confirm a diagnosis of PH.

We are also spreading the word about how people affected by certain medical conditions are more at risk of developing PH. PH can develop without any known cause (idiopathic), but it can also be linked to a number of other diseases. Connective tissue diseases like scleroderma and lupus, HIV infection, congenital heart disease, and pulmonary emboli (blood clots in the lungs) are some of the conditions that are considered risk factors for PH. We are developing partnerships with patient associations serving the communities affected by these diseases in order to reach out to people who are at risk of developing PH as a secondary condition.

In this issue of Connections, we introduce scleroderma, lupus, HIV infection, congenital heart disease, and pulmonary emboli and explain why they may lead to the development of different types of PH. We are also happy to provide information on Canadian organizations that represent patients are caregivers affected by these conditions, many of whom are also part of the Canadian PHamily. Finally, PHighters Harry Kingston, Kristen Dorscht, and Terry Anstey share their stories and talk about how scleroderma, congenital heart disease, and pulmonary emboli have been part of their PH journeys.

Connective Tissue Disease and PAH

Connective tissue diseases are systemic conditions that affect the whole body, especially joints, skin, muscles, and tendons. People living with connective tissue diseases, such as scleroderma or lupus erythematosus, can develop pulmonary arterial hypertension (PAH) as a result of damage caused by their primary condition to their respiratory and cardiovascular systems.

SCLERODERMA AND PAH

Scleroderma is a progressive chronic disease that causes hardening and thickening of the skin and internal organs. Scleroderma is an autoimmune disease, meaning that the body's immune system attacks its own tissues, resulting in inflammation and damage. In scleroderma, the body produces an excess amount of collagen (a type of protein), which can lead to scarring of the skin, tissues under the skin (including muscles and bones), and internal organs, in addition to affecting blood flow throughout the body. The factors that cause this excess production of collagen remain unknown.

Scleroderma can impact people differently, with varying degrees of severity and complications depending on whether the disease is localized (affecting areas of the skin and nearby tissues) or systemic (affecting not only the skin, but also blood vessels and major internal organs). Virtually all persons with systemic scleroderma, also known as systemic sclerosis (SSc), have some loss of lung function, often in the form of pulmonary fibrosis and/or PAH. Unfortunately, those living with SSc have an increased risk of developing PAH because of the significant fibrosis (scarring) of their lungs, which can in turn reduce blood oxygen levels and cause a reflex of increased blood pressure in the pulmonary arteries.It is estimated that 12% of all patients with systemic scleroderma develop PAH, a type of PAH know as SSc-associated PAH (SSc-PAH). Sadly, PAH is one of the leading causes of death in people living with scleroderma. Studies have also shown that SSc-PAH patients have a poorer response to therapy and significantly worse survival compared to patients with idiopathic PAH (PAH with no known cause). However, since 1996, survival rates have been on the rise thanks to treatment innovations and increased awareness of SSc-PAH.

SCLERODERMA CANADA

Scleroderma Canada | Sclérodermie Canada

Scleroderma Canada serves as an advocate nationally for those affected by the disease and works collaboratively with regional scleroderma organizations and the international scleroderma community to achieve common objectives. Scleroderma Canada is committed to promoting public awareness, supporting those affected by scleroderma, and funding research to find a cure. PHA Canada and Scleroderma Canada are partners in the Take Action PAH campaign, which advocates for PAH patients in Canada to have publicly funded access to all Health Canada approved treatments (www.takeactionpah.ca).

To learn more, visit: www.scleroderma.ca

SOURCES:

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LUPUS AND PAH

Like scleroderma, lupus is an autoimmune disease that creates inflammation in tissues and organs. The immune system of a person living with lupus attacks its own tissues, which can cause a variety of symptoms including joint pain, skin rash, fatigue, unusual reaction to sunlight, chest pain, swelling, and seizures. The specific cause of this immune system dysfunction remains unknown. Persons of all ages and genders can develop the illness, however between the ages 15 and 45, lupus occurs eight times more frequently in women then in men. It is estimated that one in one thousand Canadians are affected by lupus.

Systemic lupus erythematosus (SLE) is the most common form of lupus and can affect the skin, muscles, joints, blood and blood vessels, lungs, heart, kidneys, and the brain. Research has not clearly established a cause and effect relationship between SLE and PAH, but up to 14% of SLE patients may develop PAH as a secondary condition. This type of PAH is called systemic lupus erythematosus-associated PAH (SLE-PAH) and affects a majority of women. It is suspected that the weakening impact of lupus on blood vessels may lead to the development of PAH. Certain antibodies; elevated levels of endothelin-1, a protein produced by endothelial cells (cells lining the blood vessels); and the presence of Raynaud's phenomenon in lupus, a condition characterized by reduced blood flow to fingers and toes may also be linked to the development of SLE-PAH. Unfortunately, life expectancy for SLE-PAH patients is lower than that of idiopathic PAH patients (PAH with no known cause), but studies have shown that access to care has a positive impact on patients' survival.

LUPUS CANADA

LUPUS CANADA Life Without Lupus

Lupus Canada is a national voluntary organization dedicated to improving the lives of people affected by lupus through research, public awareness, advocacy and education.

To learn more, visit www.lupuscanada.org

SOURCES:

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HIV and PAH

Human immunodeficiency virus (HIV) is an infectious virus that weakens the immune system and can also damage other parts of the body. Without treatment, the immune system in people with HIV can become too weak to fight off serious illnesses and they can become sick with life-threatening conditions. Someone can have HIV for several years without feeling sick or experiencing any symptoms. In fact, it is estimated that 1-in-5 people living with HIV in Canada are unaware of their infection.

In 2014 there were over 65,000 people in Canada living with HIV. While the number of new HIV infections in Canada is declining, the number of people living with HIV continues to rise. In addition to new HIV infections continuing to occur (approximately seven per day in Canada), people with HIV are living longer due to advances in treatment. With proper treatment and care, most people with HIV stay healthy and live a long life.

Approximately 0.5% of people with HIV develop a type of PAH referred to as HIV-associated PAH (HIV-PAH). It is not entirely clear to researchers why PAH is associated with HIV. However, it is thought that some of the viral proteins in HIV can lead to endothelial cell injury in the lungs, as well as increased cell division of smooth muscle cells, two factors that contribute to PAH development. Even with treatment, chronic HIV can cause inflammation and immune system dysfunction, both of which may also contribute to PAH development. Treatment of HIV-PAH follows the same guidelines as idiopathic PAH (PAH with no known cause), while HIV is managed using combination antiviral therapies.

CATIE

CATIE Canada's source for HIV and hepatitis C information

CATIE is Canada's source for up-to-date, unbiased information about HIV and hepatitis C. We connect people living with HIV or hepatitis C, at-risk communities, healthcare providers, and community organizations with the knowledge, resources, and expertise to reduce transmission and improve quality of life.

To learn more, visit www.catie.ca.

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Congenital Heart Disease

PH patients and caregivers know first hand that the heart and lungs are interconnected organs. PH is a lung disease that, if not managed adequately, can impact the heart and lead to right-sided heart failure. However, pre-existing cardiac conditions, such as congenital heart disease and left-sided heart disease, can cause different types of PH.

LEFT-SIDED HEART DISEASE AND PH

Left-sided heart disease, such as left-sided congestive heart failure and mitral valve disease, can cause a type of pulmonary hypertension known as PH associated with left-sided heart disease (PH-LHD). To learn more about this type of PH, read researcher Mohamad Taha's Research Corner article on page X.

CONGENITAL HEART DISEASE AND PAH

Congenital heart disease (CHD), a condition characterized by heart defects present at birth, affects approximately 257,000 Canadians. In Canada, it is estimated that one in eighty to one hundred children are born with a heart defect, making CHD is the most common type of major birth defect. Thanks to available medical care and surgical interventions, today about 90% of children with CHD reach adulthood.

There are different types of congenital heart defects, each requiring different levels of care and intervention. In CHD, the heart's structure can present abnormalities leading to problems in passages in the heart or between blood vessels, problems with the heart valves, problems with the placement or development of blood vessels near the heart, and problems with development of the heart itself. These structural abnormalities can impact the normal flow of blood through the heart; depending on the type of defect, the blood flow can slow down, go in the wrong direction or to the wrong part of the heart, or be blocked.

Some CHD defects allow blood from the left-side of the heart to leak to the right-side of the heart. This leak increases the blood flow through the pulmonary arteries and lungs, which over time can lead to the development of pulmonary arterial hypertension (PAH). This type of PAH, known as congenital heart disease associated PAH (CHD-PAH), is also referred to as Eisenmenger syndrome. Examples of congenital heart diseases that can lead to PAH include atrial septal defect (ASD) and ventricular defect (VSD). The heart of person with ASD presents an abnormal opening in the wall between the left and right atriums, the chambers located in the upper portion of the heart. The heart of person with VSD presents an abnormal opening in the wall between the left and right ventricles, the chambers located in the lower portion of the heart. In both cases, blood leaks between the left and right sides of the heart, increasing blood flow through the lungs, which in turn increases pulmonary arterial pressures.

CANADIAN CONGENITAL HEART ALLIANCE

The Canadian Congenital Heart Alliance's mission is to improve the quality of life and health outcomes for individuals with congenital heart defects by raising awareness, providing peer support and mentoring, advocacy, and advancing research.

To learn more, visit www.cchaforlife.org.

SOURCES:

"About Congenital Heart Defects," Canadian Congenital Heart Alliance, http://www.cchaforlife.org/about-congenital-heart-defects.

"Can Heart Disease Cause PH?," LivingwithPH.ca, Pulmonary Hypertension Association of Canada, http://livingwithph.ca/en/disease/can-heart-disease-cause-ph/.

"Facts and Issues," Canadian Congenital Heart Alliance, http://www.cchaforlife.org/facts-issues.

Pulmonary embolism (PE) refers to blockage in the pulmonary arteries, most commonly caused by blood clots. PE can lead to the development of a type of PH called chronic thromboembolic pulmonary hypertension (CTEPH). Early detection of CTEPH is crucial because it is the only type of PH that has the potential to be cured through surgery.

Pulmonary Embolism and CTEPH

Pulmonary embolism (PE) occurs when blood clots (thrombosis) develop in the body, travel to the lungs, and get lodged in the pulmonary arteries. In some cases PE can develop without any known cause. However, in most cases, PE is caused by a condition called deep venous thrombosis (DVT), which occurs when abnormal blood clots develop in large veins, usually in the leg or pelvis. The clots can break off and travel to the lungs, causing PE. Risk factors for DVT include immobility (e.g. being in bed for days or taking a flight over six hours long); injury to blood vessels due to broken bones or surgeries; and hypercoagulability (increased risk of clotting) due to medical conditions, hormones, or genetic factors. Pulmonary embolism and DVT are types of venous thromboembolism (VTE). VTE is a common condition that affects approximately one in 1,000 persons each year.

People with pulmonary embolism (PE) are at higher risk of developing chronic thromboembolic pulmonary hypertension (CTEPH) if the blood clots in their pulmonary arteries do not dissolve. Pulmonary embolism can be acute or chronic. In acute PE, the clots can be dissolved with an anticoagulation treatment (blood thinners). If the clots do not dissolve completely, PE becomes chronic. Chronic PE can lead to the formation of scar tissue in the pulmonary arteries, which can cause CTEPH. Up to 4% of patients may develop CTEPH within two years following PE. Unlike other types of PH, CTEPH has the potential to be cured through the surgical removal of the clots and scar tissue blocking the pulmonary arteries. For more information on the surgical treatment of CTEPH, read Nurse Practioner Anastasia Bykova's article, *"Patient Journey in Chronic Thromboembolic Pulmonary Hypertension (CTEPH),"* pages 24-25.

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Thrombosis CanadaTM is dedicated to furthering education and research in the prevention and treatment of thrombotic vascular disease.

~~For more information,~~ visit www.thrombosiscanada.ca.

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Thrombosis Canada, You Have a Deep Vein Thrombosis (DVT), Disease Information Sheet, http://thrombosiscanada.ca/wp-content/uploads/2016/10/M159-You-Have-a-DVT_Oct2016.pdf.

Being and Staying in Command: Harry Kingston's Journey

To provide insight into my personality, I am a senior National Security Advisor with a Military Officer background, temperament, attitude, and drive. This experience and training have enabled me to cope with the different challenges that have come my way. "Never surrender" is a phrase I live by. I further believe that one cannot make excuses in life; you simply deal with whatever you have been given. However, since receiving my pulmonary arterial hypertension (PAH) diagnosis in 2012, these convictions have been put to the test.

I live with both scleroderma and PAH. To date, I have been moderately impacted by scleroderma, which I manage well. Associated symptoms include skin-related issues and a consistent iron deficiency, for which I receive a monthly IV supplement. My PAH is much more difficult to manage; my care has involved complex medical procedures and has caused me the most stress and lifestyle challenges. At the moment, my medical team is concerned with adjusting my Caripul IV treatment, which I have been on since July 2016, and constantly dealing with effects of the progression of right-sided heart failure. I believe that each of us must trust our medical team of specialists. In my experience, following my medical team's advice, which can sometimes be blunt, is absolutely critical. I trust in the great team that follows me at the Ottawa PH Clinic, as well as in my nephrologist and scleroderma specialist.

I pride myself in being active in all aspects of life, no matter what. Given my PAH, stress that accompanies my professional position can be very draining. However, my biggest concern is the possibility of not being able to access the treatment or combination of treatments that I may need in the future. Having this fear simmer on the backburner of my mind places further strain on my energy and physical condition, especially because I believe it is caused by politics and the government refusing to change its mind.

Thankfully, my amazing wife, Teri, a PHA Canada Ambassador, helps me get through very difficult days. Together, we rely on our faith and we count on our church family to provide payer and emotional support. I have found that most people

"To encourage my fellow PHamily, one of the most important things I think people living with PH can do is become "trailblazers". We can all benefit and learn from each other's personal journeys as we collectively build upon an increasing body of patient-based knowledge."

are very supportive when they learn of my condition. However, like most, Teri and I have also encountered people with no concept of chronic suffering, personal pain, or commitment to duty over self. Members of the Ottawa PH Support Group have been a major relief to us. We became involved immediately after my initial diagnosis, something I would recommend to every PH patient. Our group forms a welcoming and mutually supportive community; our monthly meetings are opportunities to get energized and encourage others.

To encourage my fellow PHamily, one of the most important things I think people living with PH can do is become "trailblazers". We can all benefit and learn from each other's personal journeys as we collectively build upon an increasing body of patient-based knowledge. We can all learn from shared experiences about living with PH in all its variations. The best advice I would give to a patient newly impacted by this disease is to live each day with the same passion one feels when they are burning to tell that special someone just how much they care about them! You do not have a lot of time for pity or regret.

My whole life has been focused on service, duty, and commitment to servant leadership a philosophy in which a citizen's primary role is to reach out to serve others. Joining PHA Canada's Board of Directors in 2013 therefore came as a natural extension of my commitment to being a positive force in our community. I know first-hand that the support provided by PHA Canada is critical for many patients and families and I want to give back while I can. In my opinion, the most important role that our organization plays at the moment is advocating on behalf of the whole PH community, especially when it comes to fighting for access to treatments.

PHA Canada gives me hope for the future. I truly think we will see tremendous progress in access to and development of new treatments in the years to come. I pray and do as much as I can for the continued wellbeing of our community. As a Board Member, my goal is to ensure PHA Canada's sustainability and continued growth. On a personal level, my hope for the near future is to finally transition from my professional career so that Teri and can focus on our dreams, like seeing one of our kids get married. I PHight for that every day.

Contributed by: Harry Kingston, scleroderma and PAH patient and PHA Canada Board Member

The Dorscht Family doesn't let PH get in the way of family fun.

A Heart of Gold: The Dorscht Family's Story

Ella was born with a heart defect known as transposition of the great arteries (TGA). TGA can be treated with open-heart surgery in the first few days of life, which usually entails a fairly straightforward recovery, but Ella had a string of complications after her TGA switch operation and required more surgeries. After a second open-heart surgery to fix an aneurysm, Ella's heart would not pump. She was put on extracorporeal membrane oxygenation (ECMO), a form of life support, for four days. After she was on ECMO, Ella outwardly seemed to make a quick recovery, but her echocardiograms showed something was still wrong. Ella had a catheter surgery to try to pinpoint the problem, which is when she was diagnosed with PH, just shy of 6 months old. She did not have any symptoms of PH other than what the heart tests showed.

Ella is only three years old but she has already had two open-heart surgeries, six heart catheter surgeries, and a chest debridement surgery. Now, she takes daily oral medications and uses oxygen 24/7, even though her oxygen level is high, to help lower her lung pressures. It is always interesting to explain to a new doctor why a child with oxygen saturations of 99% requires oxygen!

Having a very active preschooler attached to oxygen 24/7 is difficult. When Ella started using oxygen three years ago, she couldn't even sit. Now she never sits still! She often gets her tubing caught on furniture and doors, and needs someone to follow her every step while out and about at the playground, water slides, you name it. Thankfully, Ella's big big brother, Cameron, is a great helper and is very patient with his sister.

Through each of Ella's developments stages baby, toddler, and now preschooler my husband, Nick, and I have definitely learned a lot about managing oxygen. Every new development has posed new issues. For example: "Now that she can roll, how do we prevent her oxygen from wrapping around her in her crib?" Through lots of trial and error, we ~~came~~ with solutions, and, as there is little to no information available, ~~share~~ our tips with other heart and PH parents. Overall, what we have learned is that, while your child's diagnosis may absolutely come as a surprise, it doesn't have to define your lives. Ella does many things typical children do: she swims, goes to music class, went on an airplane, etc. Most things need a little bit of thought or modification, but they can be done.

For example, our family had a magical experience when we went to Disney World last year. We had a day filled with a chain of extra good things that Disney employees call "pixie dust." No one knew of Ella's diagnosis, but somehow she managed to charm her way into a whole day of wonderful surprises. We were treated to lunch, her brother Cameron rode a ride with the Mad Hatter, and the kids got to meet seven Disney characters without waiting in line! It was amazing to have Ella treated to such a great day. After all she has been through, it was such a well-deserved reward that just seemed to happen by magic!

Our family is now spreading a bit of magic closer to home. After Ella was born, my husband and I stayed for two months at the Toronto Ronald McDonald House and attended many Home for Dinner events home-cooked meals for families of sick children put on by corporations. We both agreed

"Ella does many things typical children do: she swims, goes to music class, went on an airplane, etc. Most things need a little bit of thought or modification, but they can be done."

this was by far the "best" part of a very difficult hospital stay and vowed to give back when Ella came home. If a corporation could do it, why couldn't we? I am proud to say that we will be hosting our fourth Team Ella Home for Dinner this year! We couldn't do it without an amazing group of friends and family who help with everything from fundraising to cooking and serving. Ella also received well over fifty units of blood as an infant and Nick and I said we wanted to put back into the system what Ella had used. For the past two and a half years, we have held Team Ella Blood Drives in our community. So far, Team Ella has donated close to two-hundred-and-fifty units of blood, so we have definitely surpassed our goal!

Contributed by: Kristen Dorscht, PH caregiver, Elmira, ON

On the Road to Overcome CTEPH: Terry Anstey's Story

Thanks to his PEA surgery, Terry Anstey is able to lead an active lifestyle.

I started to experience shortness of breath, general fatigue, faintness, exercise intolerance, and swelling of the feet (pedal edema) in early 2010 while on an overseas work assignment. My GP suspected a mild heart attack and referred me to an internal medicine specialist who ordered various tests, including pulmonary function tests and an echocardiogram. The verdict was chronic obstructive pulmonary disease (COPD). I was prescribed different inhalers over the ensuing months, but none had noticeable beneficial effect. I was also advised to eat less salt to help with my edema.

Despite months of this treatment, my condition deteriorated and my GP referred me to a Breath Program at Lions Gate Hospital (LGH) in North Vancouver, BC. In the fall of 2011, to register for this program, I met LGH's respiratory specialist. Within short order, he suspected I had a "potentially life-threatening condition" and admitted me to LGH that same day for observation and tests. Following a week of testing, he concluded that my symptoms were inconsistent with COPD (although I do have moderate COPD) and diagnosed chronic thromboembolic pulmonary hypertension (CTEPH). I was prescribed anti-coagulation tablets (Warfarin) and oxygen and was referred to Dr. John Swiston at Vancouver General Hospital's PH clinic. Dr. Swiston confirmed my CTEPH diagnosis in January 2012, two years after the appearance of my symptoms.

Like most people, I had never heard of PH before being diagnosed with it. I did not have the medical knowledge to connect my symptoms to this disease and had no history of blood clots before receiving my diagnosis although, obviously, I must have had one since CTEPH is caused by blood clots migrating to the lungs. Looking back, my lifestyle might have been conducive to blood clotting. Sitting for long periods of time is a risk factor for the development of blood clots and, through my work, I was travelling about 100,000 miles of long-haul flights per year.

In April 2012, results from a right-heart catheterization indicated that my heart was overstrained. The Vancouver PH clinic's medical team advised me that, without treatment, statistical life expectancy after diagnosis was about two years, that drugs were available to slow disease progression, but only surgery could "cure" CTEPH. However, I was told that, due to results from chest radiology imaging and my age (then 68 years), I was not a candidate for pulmonary endarterectomy (PEA) surgery. I was prescribed Tracleer® and increased oxygen. A year later, in June 2013, the Vancouver clinic referred me to the Toronto CTEPH clinic for reassessment. I had a teleconference with Dr. Marc de Perrot, the clinic Director, and was accepted for surgery subject to tests.

I travelled to Toronto with my wife and was admitted to Toronto General Hospital for surgery on September 14, 2013. I had my surgery two days later. Post-surgery, I spent about one week in critical care and two weeks in general care before being discharged from hospital. Lying in a hospital bed for 3 weeks was, of course, frustrating, but the nurses were very tolerant when I tried to get up before I was strong enough and ended on the floor. Daily assisted exercise helped me get strong enough to be discharged and I flew home to Vancouver on October 6, 2013.

PEA surgery is not a "cure" per se, insofar as blood clots in the smaller pulmonary vessels cannot be removed surgically. However, having blood clots and scar tissue removed from my main pulmonary arteries has had a huge impact on my symptoms. Within just 3 weeks of surgery, tests indicated that my right-heart function had returned to normal. I no longer use oxygen or inhalers and can do most things, albeit more slowly than I used to. I still get out-of-breath upon overexertion, but I maintain an active lifestyle. Prior to surgery my condition had deteriorated to the point of not being able to walk more than 25m without rest. Now I walk about 5km a day, at a pace of about 4km/h, and I'm travelling internationally again.

Some people, upon learning details of the surgical procedure (including full cardio and pulmonary suspension), might be apprehensive to proceed. For me, however, the decision was an easy one, considering my unacceptable quality of life and the shortened life expectancy I faced without the surgery. I have since recommended the procedure to others.

I recently shared my story in PHA Canada's PH Awareness video and was pleased to have that opportunity to raise awareness both within the medical profession and the general public. One problem with PH generally, and CTEPH in particular, is that it is a rare disease that shares many of its symptoms with other, more common diseases, with the result that many medical practitioners fail to diagnose it. Failure to diagnose, misdiagnosis, or delayed diagnosis will have detrimental effects. On a social level, another problem is that persons with PH don't look ill might they be suspected of malingering? PH patients may not want sympathy, but surely deserve understanding.

Contributed by: Terry Anstey, CTEPH patient, North Vancouver, BC

CTEPH

Chronic **T**hromboembolic **P**ulmonary **H**ypertension

Patient Journey in Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

Anastasia Bykova, Nurse Practitioner

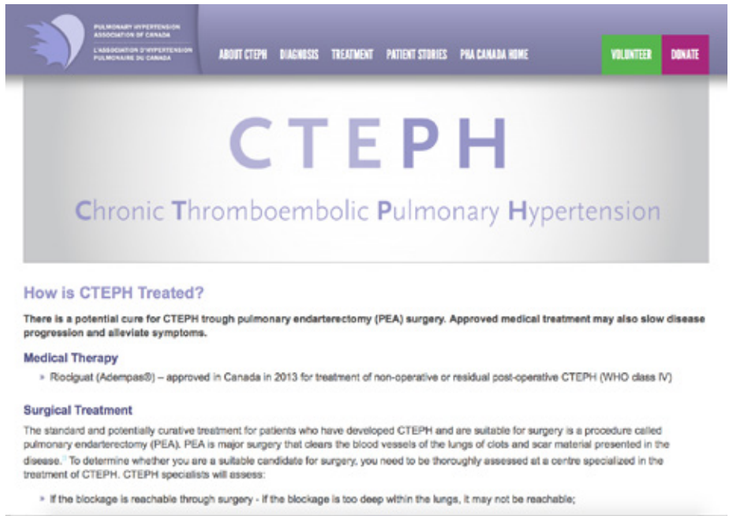
In the Spring 2016 issue of Connections, I published an article explaining how chronic thromboembolic pulmonary hypertension (CTEPH) develops and is treated. In this follow-up article, I will talk about the patient experience of being diagnosed with CTEPH and treated through pulmonary endarterectomy (PEA surgery). Given the complexity of the disease process and the time it takes for an accurate diagnosis of CTEPH to be established, patients and their families share a lengthy and emotionally challenging journey.

THE BEGINNING ONSET OF SYMPTOMS

CTEPH remains an underdiagnosed disease in which chronic pulmonary emboli (blood clots) obstruct the pulmonary arteries. This process leads to elevation of blood pressure in the pulmonary arteries, causing pulmonary hypertension (PH) and stress on the right side of the heart. Initially, patients develop shortness of breath (SOB), which is mainly noticeable when navigating inclined surfaces (i.e. climbing stairs, going uphill) or with strenuous physical activity (i.e. gym, sports, mowing the lawn). As the disease progresses, patients experience fatigue and breathlessness even when travelling on flat surfaces (i.e. walking in a mall or on a street). Patients will commonly presume that they are "getting out of shape" to explain these limitations. In reality, it is the underlying CTEPH that limits their exercise ability and impacts their fitness level. In an effort to get in shape, many people try to engage in physical activity while still experiencing progressive SOB and fatigue. Eventually, they will give up their efforts and over time, adapt to these symptoms by doing less exercise and adopting a more sedentary lifestyle. This adaptation process leads to weight gain and physical deconditioning, which in turn exacerbates symptoms of SOB and fatigue. With time, some people may develop additional symptoms such as chest pain, fainting, and swelling of the legs and abdomen, and may require home oxygen to maintain their oxygen level. This vicious cycle may go on until an accurate diagnosis is established.

CTEPH DIAGNOSIS AND TREATMENT

CTEPH symptoms are non-specific and can be associated with a number of more common medical conditions. As a result, the majority of CTEPH patients will be seen by multiple specialists before a definitive diagnosis is established. Patients and their families generally find the process of going from one specialist to another in search of a final diagnosis to be frustrating. The average time from the onset of symptoms to CTEPH diagnosis is 14 months, which demonstrates the difficulty of establishing an accurate diagnosis. The process may go as follows: a respirologist may treat a patient whose CTEPH has not yet been diagnosed with antibiotics and "puffers". When the symptoms do not improve, the patient will be referred to a cardiologist for more investigations. Eventually, a PH program will evaluate the patient and confirm a CTEPH diagnosis. Importantly, whether CTEPH is confirmed or suspected, all patients should be assessed for poten-



Thanks to a collaboration with the University Health Network and the Toronto CTEPH clinic, we have published new educational resources on our redesigned CTEPH website. Visit www.phacanada.ca/cteph to access information and resources.

tially curative pulmonary endarterectomy (PEA surgery) by a program specialized in the treatment of CTEPH. Early CTEPH diagnosis and referral for surgical evaluation are crucial to improve post-operative recovery and increase the chance of a cure.

BEFORE AND AFTER PULMONARY ENDARTERECTOMY (PEA SURGERY)

PEA surgery is a gold standard treatment for CTEPH. In Canada, expertise in PEA surgery is regionalized, which allows the formation of strong, multidisciplinary teams of experts who can provide highly specialized patient care. For many patients and their families, this means travelling to Toronto or Ottawa to receive a potentially curative treatment.

BEFORE PULMONARY ENDARTERECTOMY SURGERY

The Toronto CTEPH Program is the largest Pulmonary Endarterectomy Program in Canada and gets referrals from across the country. Once a CTEPH diagnosis has been confirmed and PEA surgery in Toronto is planned, the majority of patients and their families feel overwhelmed. The most common reasons are 1) the disease process and surgical procedure are complicated to understand; and 2) the trip to Toronto takes a lot of preparation. It is important to let CTEPH patients and families know that they are not alone in this process, and that they can reach out to their CTEPH team for guidance. The key is for the family members to help a patient prepare for the trip by working collaboratively with the CTEPH team. Educational materials that the medical team will provide on CTEPH diagnosis and PEA surgery can alleviate some anxiety for patients and families. An information package with clear instructions and a checklist will be sent out to ensure that patients and family members know how to prepare ahead of time. Some of the items patients and families need to plan for include: 1) identifying a support person who will accompany the patient home after their hospital discharge; 2) securing provincial health coverage/travel and accommodation assistance (if applicable); and 3) setting up home oxygen for the flight and stay in Toronto. The more prepared the patient and family will be, the less overwhelmed and stressed they will feel away from their home.

AFTER PULMONARY ENDARTERECTOMY SURGERY

The majority of patients report an immediate and ongoing improvement of SOB following PEA surgery. If the chronic clots were in the smaller pulmonary arteries, causing less SOB symptoms, the breathing will improve at a slower rate over the next few months. Families may notice initially that some patients demonstrate subtle personality changes, confusion, or even inability to focus (i.e. taking twenty minutes to write an email that would have taken two minutes to write before PEA). These mental changes are temporary and will resolve within the first few weeks as the patient recovers. One third of the patients will require supplemental home oxygen following PEA surgery even if they were not on oxygen before. This is not a reason to be alarmed that the surgery did not work! The newly unobstructed pulmonary arteries will relearn to control the amount of blood flow to match the air moving in and out of the lungs and the patients will be weaned off home oxygen within a few weeks or months (depending on how sick they were before surgery).

The key is to be patient with the body and give it the necessary time to heal. As the body heals, the patient will notice significant improvement in breathing, resolution of fatigue, increased ability to be active and exercise, and sharper mental function due to the increased amount of oxygen that is available to the brain. Going through PEA surgery is a challenging process, but with proper preparation and support from a dedicated surgical team, patients and their families can confidently embark on this journey to significant quality of life improvements and a potential cure to CTEPH.

For more information on CTEPH, including contact information of clinics specialized in the treatment of CTEPH in Canada, and to read Anastasia Bykova's article, "CTEPH: Curable Type of Pulmonary Hypertension" (Connections, Spring 2016. Vol. 7, No. 1), visit www.phacanada.ca/cteph.

Contributed by: Anastasia Bykova, Nurse Practitioner, Toronto Chronic Thromboembolic Pulmonary Hypertension Program

Meet Your Medical Professional: Dr. Marc de Perrot

Dr. Marc de Perrot, MD, MSc, is the Director of the Toronto Chronic Thromboembolic Pulmonary Hypertension (CTEPH) Program at the University Health Network's Thoracic Surgery Clinic. A leader in the treatment of CTEPH in Canada, Dr. de Perrot has changed the lives of many CTEPH patients by performing pulmonary endarterectomy (PEA surgery), a procedure that has the potential to cure this particular type of PH. Dr. de Perrot also occupies the position of Associate Professor, Division of Thoracic Surgery, at the University of Toronto, is affiliated with the Toronto General Research Institute (TGRI), and is a member of PHA Canada's Medical Advisory Committee (MAC). We are pleased to share insight into his medical practice through this interview.

PHA Canada: *Where did you begin your medical career, and in what field?*

Dr. de Perrot: I started my training in general surgery in Geneva, Switzerland and then came to Toronto to complete my training in thoracic surgery and lung transplantation.

PHA Canada: *How were you first introduced to pulmonary hypertension? What drew you towards developing a specialization in the field of PH, and in the treatment of CTEPH more specifically?*

Dr. de Perrot: My initial exposure to pulmonary hypertension was through the Toronto General Hospital's Lung Transplantation program in the early 2000s. At that time, patients with CTEPH were still referred for lung transplantation. This encouraged me to spend an extra year of training in Paris, France to learn more about CTEPH and acquire the necessary skills to start a Pulmonary Endarterectomy (PEA surgery) Program in Toronto as an alternative to lung transplantation.

PHA Canada: *CTEPH is a form of PH that is potentially curable through PEA surgery. What has been your experience performing this surgery in terms of improving patients' lives?*

Dr. de Perrot: This is a unique surgery in the sense that you truly can transform a patient's life, both in terms of quality and expectancy. As I mentioned, in the past lung transplantation was being performed for patients with CTEPH, leading to a lot of new medical challenges related to immunosuppressive medications. In contrast, after pulmonary endarterectomy, patients' PH may be cured and they may be able to resume their normal life, while only taking an anticoagulation treatment in the long term. This is unique in the field of end-stage lung disease and pulmonary hypertension.

PHA Canada: *Can you share some inspiring stories of CTEPH patients whom you successfully treated?*

Dr. de Perrot: Every patient who proceeds to pulmonary endarterectomy is inspiring and has a personal story that would be worthwhile sharing. It is always fascinating to see the patient's transformation from before to after the surgery.

PHA Canada: *Working in the field of PH, what do you find most challenging? Are there particular challenges associated with treating CTEPH?*

Dr. de Perrot: Pulmonary hypertension is a challenging field due to the nature of the disease. This is primarily a disease of the lungs, but it can progressively damage the heart, liver, and kidneys, until eventually every organ in the body is affected by the disease. Our goal in treating CTEPH is to detect the disease before the heart and other organs become damaged. When pulmonary endarterectomy is performed early on in the course of the disease, you can minimize the risks of the surgery and maximize chances to normalize the condition.

PHA Canada: *You are a member of PHA Canada's Medical Advisory Committee (MAC). Can you explain why you became involved with PHA Canada?*

Dr. de Perrot: PHA Canada is a very important organization in the country. It is a platform that is accessible across Canada to all patients and their loved ones, allowing them to connect with each other and share their experiences. PHA Canada also plays a unique role in ensuring that up-to-date quality medical information is available to patients. This organization empowers patients to take an active role in their treatment and ensure that they have access to the care they need.

PHA Canada: *Are you currently involved in research or clinical practice development projects?*

Dr. de Perrot: The CTEPH Program's team has several ongoing projects both in clinical care and in the laboratory. Our research is oriented towards diagnosis and treatment. More specifically, we are looking at the impact of PH on the metabolism to diagnose the disease at an earlier stage and be able to make better predictions by understanding how PH affects the heart.

PHA Canada: *What is the most important information would you want to share with someone who is newly diagnosed or newly affected by CTEPH?*

Dr. de Perrot: CTEPH is the only type of pulmonary hypertension that can potentially be cured. It is therefore important to search for the disease in any patient with a new diagnosis of PH. A ventilation-perfusion scan (VQ scan) remains the best test to screen for CTEPH and should be done in all patients with PH.

PHA Canada: *What advances have you seen in the treatment of CTEPH since you started practicing? What do you find to be the most encouraging research/developments currently taking place?*

Dr. de Perrot: The field of CTEPH has changed a lot over the past 15 years. Pulmonary endarterectomy has become a safe surgery that can be performed in more than 80% of patients with CTEPH. Specific medication such as riociguat has also become available for CTEPH patients who are not candidates for surgery or with residual PH after surgery. More recently, we have started a program of balloon pulmonary angioplasty (BPA) as an additional option for some patients with CTEPH. I think the most encouraging development is that we are finally providing optimal medical care to a large number of patients who were previously unaware of this condition. There is however a lot of more work to do as CTEPH is still largely underdiagnosed in Canada.

Contributed by Dr. Marc de Perrot, MD, MSc, Director, Toronto Chronic Thromboembolic Pulmonary Hypertension (CTEPH) Program at the University Health Network's Thoracic Surgery Clinic

Mohamad Taha is a PhD student and PH researcher under the supervision of Dr. Duncan Stewart at the University of Ottawa. Mohamad contributes a bi-monthly Research Corner to our Pulse e-newsletter. ~~We are glad to have Mohamad's contribution to provide our community with insight into the PH research process.~~

Pulmonary Hypertension Associated with Left-Sided Heart Disease (PH-LHD)

In this edition of *Research Corner*, we will address questions regarding pulmonary hypertension associated with left-sided heart disease (PH-LHD), which is classified as Group II pulmonary hypertension.

What is pulmonary hypertension associated with left-sided heart disease (PH-LHD) and how does it develop?

To understand PH-LHD, we must know how our heart and lungs interact. Different sides of our heart are responsible for pumping blood through our lungs, where it is oxygenated, and then through the rest of our body. After our body uses the oxygen in our blood, the de-oxygenated blood passes through the right side of the heart (right ventricle), which pumps it into the pulmonary artery and then into the lungs. This usually happens at a relatively low pressure in the right ventricle and pulmonary artery. Blood is re-oxygenated in the lungs and then passes into the left side of the heart (left ventricle), which pumps it out into the rest of the body. This process takes place at pressures five times higher to allow for proper delivery to the extremities.

People suffering from left-sided heart disease can develop higher pressures in the left-heart or dysfunction of the heart valves, whose role is to prevent the blood from going backward into the lungs. This high pressure and/or valvular dysfunction (dysfunction of the heart valves) results in blood being pushed back into the lungs. This damages the blood vessels in the lungs and creates more resistance for the blood flowing in the proper direction, eventually leading to high pulmonary pressures and PH.

How prevalent is PH-LHD?

Due to the high prevalence of left-sided heart disease (LHD), PH-LHD is the most prevalent form of PH. It's very difficult to get exact numbers on its prevalence due to the variety of types of left-sided heart disease that can lead to PH. Some LHD patients are prone to the development of PH-LHD while others aren't.

How does the treatment strategy for PH-LHD differ from PAH?

Many of the therapeutic interventions targeting PH-LHD focus mainly on managing the underlying left-sided heart disease. Studies using PAH therapies did not show much improvement in PH-LHD patients. This is most likely due to the different origins of the two diseases; whereas PAH develops in lung arteries, PH-LHD develops in lung veins. However, recent studies using sildenafil (a PDE5 inhibitor) in specific subgroups of PH-LHD have shown some promise. Research continues to be conducted in order to find novel therapies for PH-LHD.

References: Georgiopoulou et al. (2013), "Left Ventricular Dysfunction With Pulmonary Hypertension, Part 1: Epidemiology, Pathophysiology, and Definitions." Circ Heart Fail; 6:344-354.

Kalogeropoulos et al (2013), "Left Ventricular Dysfunction With Pulmonary Hypertension,

Part 2: Prognosis, Non Invasive Evaluation, Treatment, and Future Research." Circ Heart Fail; 6:584-593

Please always keep in mind that while I can provide you with a small insight into PH research, you should always be able to get answers from your pulmonary hypertension Specialist, who is more familiar with your specific case and your treatment history.

Contributed by: Mohamad Taha, PhD Candidate under the supervision of Dr. Duncan J. Stewart, Department of Cellular and Molecular Medicine, Faculty of Medicine, University of Ottawa.

In Memory 2016

While our community holds enormous hope for the future, the reality is that pulmonary hypertension still takes loved ones away from us. In 2016, our community has sadly lost the PHighters listed below. Our hearts go out to their families and friends.

Keith Brooks
Mélissa Cadorette
Jenna Noelle Comeau
Susan Crowder
Evelyn Dragojevich
Edward Earl
Susan Garland
Desmond Hollingberry
Merlin Jarmain
Florence Kelly
Guy Laroche
Francine Lavoie
Manon Lavoie
Helen Lyons
Quentin Mosiondz
Rose Nicoletti-Shreve
Lorna Pennings
Lucia Petta
Hélène Provost
Dolores Rapinchuk
Sydney Roberts
Carolyn Robinson
Paciencia Santos
Janessa Siebert
Wayne Smalldon
Christina Thorn
Merle Wolfe

2016 Donor Recognition

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PHA Canada's Corporate Committee members pay annual dues in addition to providing sponsorships in support of our programs and services. Our 2016 Corporate partners were:

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Pfizer Canada (Bronze level sponsor)
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United Therapeutics Corporation (Gold level sponsor)

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PHA Canada relies on the generosity and dedication of our donors to further our mission and to support our activities. We would like to thank everyone who contributed in any way in 2016. We appreciate each gift, as together they make a huge difference in the lives of those living with PH. We would like to particularly recognize and thank the donors below who personally contributed donations of over $500.

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Quentin Smith

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WE WOULD ALSO LIKE TO RECOGNIZE AND THANK ALL THOSE WHO ORGANIZED AND CONTRIBUTED TO THE FUNDRAISING EVENTS LISTED BELOW:

6-Minute Walk for Breath *(Ottawa, ON)*
20 Mile Longwoods March in Honour of Everleigh Pierce *(London, ON)*
Ascension du Mont-Washington *(Mount Washington, NH)*
Be Brave and Sparkle *(Newmarket, ON)*
Brooklin Quiz Night *(Brooklin, ON)*
Fourth Annual Dine for the Cause *(Ottawa, ON)*
Fourth Annual Masquerade Ball *(Calgary, AB)*
GolPH for PH *(Brampton, ON)*
Lend a Hand for PH 2016
Music for Mia: A Concert for PH *(Bradford, ON)*
Personal Pages for PH
Quentin's PHight for PH Awareness *(Thompson, MB)*
Sail-o-thon Benefitting PHA Canada *(Vancouver, BC)*
Samantha Bowker's PH Run/Walk *(Victoria, BC)*
Third Annual Run/Walk for PH Research *(Ajax, ON)*
Third Annual Run/Walk, Swim, or Bike/Spin PH Challenge *(Bradford, ON)*
Volleyball Tournament in Memory of Brenden Brinkworth *(Long Sault, ON)*

Connections submissions guidelines

The deadline for submissions for the next issue of ***Connections*** is November 15th, 2016. ***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. We'll accept articles, personal PH stories, quotes, photos, tributes, poems, drawings and more for publication in the magazine. 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 about!

Please send submissions including your contact information to:

Mariane Bourcheix-Laporte
connections@phacanada.ca
Subject: Connections submission

We look forward to reading your stories!

Work submitted will be printed as space permits.

Imagine Canada Standards Accreditation

In May 2015, PHA Canada became accredited as part of the Imagine Canada Standards Program. This is a big step for PHA Canada who has now joined the ranks of Canada's most trusted charities and non-profits! To receive this accreditation, organizations must demonstrate excellence in the areas of Board Governance, Financial Accountability and Transparency, Fundraising, Staff Management and Volunteer Involvement. To learn more about this accreditation, visit **phacanada.ca/accountability**.

Connections 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.