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

# CONNECTIONS

**The Official Magazine of the Canadian PH Community**

CONNECTIONS | Spring 2016 | *Vol. 7, No. 1*

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All of the photographs in this issue were contributed by the article authors or by PHA Canada.

# Message from the Chair: The Winds of Change

Hello PHriends,

As most of you have no doubt heard, PHA Canada has gone through tremendous changes over the past few months. We have new staff in the office and a new direction! Very excitingly, Ms. Jamie Myrah has joined PHA Canada as our first-ever Executive Director! Jamie brings a wealth of experience in the health charity field, as well as tremendous energy and passion to help PHA Canada grow in order to better support our vision of a better life for all Canadians affected by PH.

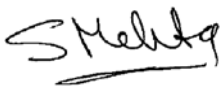
There are other news affecting the Canadian PH community. Bad news. Macitentan (Opsumit), the newest oral medication approved by Health Canada for the treatment of PAH has been denied public government funding by the pan-Canadian Pharmaceutical Alliance (pCPA), and thus, will not be available to most Canadian PH patients. Macitentan was approved for public funding in Quebec in 2013, and has been available to treat Quebec PAH patients since then. Moreover, there are private insurance companies that are already paying for macitentan for some PAH patients across Canada. However, for the majority of non-Quebec Canadian PAH patients, the recent pCPA decision makes it so that macitentan will not be available to them as a treatment option, even if their PH expert physician decides it may be the best medication for them.

Although Canada does have a national "universal" healthcare system, this does not mean that all Canadians have access to the same excellent care and treatments for PH. Indeed, each individual provincial/territorial Ministry of Health makes their own decisions as to which PH treatments are funded for which PH patients. Moreover, some recent government decisions mandate in what order PH medications may be used, i.e. patients only have access to drug "A" first (often the cheapest one), and may have access to drug "B" (even if it may be more effective than drug A) only in case of a lack of improvement or actual worsening. As such, it is government bureaucrats that will often decide which therapy may be most appropriate for a PH patient. Clearly, this is a treatment approach that is not only inappropriate, but dangerous for many patients with a serious, usually progressive, and often fatal illness like PH. I think we all agree that the best treatment for each individual Canadian PH patient can only be decided by their expert PH physician.

Since PHA Canada was founded, one of our most important concerns has been working towards ensuring access for all Canadian PH patients to all effective Health Canada-approved PH therapies, regardless of patients' financial resources, private insurance coverage, or province/territory of residence. Indeed, advocacy on behalf of the Canadian PH community is one of the 5 key priorities in our Strategic Plan. Such efforts have been ongoing, as described in the current issue of Connections, and will become even more of a priority in 2016 and beyond.

To fight such restrictions on PH patients' access to all effective PH therapies, PHA Canada needs your help. Government, media, and other community organizations are often most responsive to the individual, powerful voices of those directly affected by an illness. Individuals like you—PH patients and caregivers—can have a tremendous impact by sharing your stories of living with PH—the frustrations, fears, sadness, but also the victories and especially the hope—with PHA Canada, the media, provincial representatives, and the public. Help those living with PH across Canada by speaking out and getting involved in our ongoing advocacy campaigns.

With hope,



Dr. Sanjay Mehta

PHA Canada Board Chair

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# CONNECTIONS

## *In This Issue:*

# Sock Monkeys, Chest Tubes and Physiotherapy: How I Got my New Lungs

*Amanda holding a sock monkey. While in the hospital, she and her children had the same sock monkeys to comfort them.*

All of a sudden, at 3:30 a.m. on Saturday October 17, 2015, the light in my hospital room is switched on. A doc whom I've never met says: "I don't want you to eat or drink anything because we might have lungs for you much later today. Now try to get some rest." Then he leaves. Rest... Sure. I lay there quietly, and said a quiet thank you to a family, somewhere, who had just lost their loved one and made the decision to move ahead with organ donation.

I had been admitted to Toronto General Hospital on August 10 for fluid retention related to my IPAH and only returned home months later. My medication was not working well for me. My heart was failing. Other organs were stressed. One of the hardest things about being hospitalized was being an hour away from my husband and three young children. We scrambled to find summer camps and after school care for them. My amazing husband continued to work full time and took care of everything at home. It is difficult to parent from a hospital bed, but we endured and tried to make the best of it: we celebrated my daughter Grace's 9th birthday in the hospital, I shopped for back to school stuff online, my sister and my parents took the kids out, and I had a day pass for Thanksgiving! During my hospitalization, I struggled through two painful subcutaneous Remodulin site changes and was severely restricted with fluids (it felt like I was on a desert island...). Through all of the ups and downs I never lost hope that my PHight was not over. I was confident that the best was yet to come.

***Through all of the ups and downs I never lost hope that my PHight was not over. I was confident that the best was yet to come.***

And then, my day came. The troops mobilised and family and friends were at the hospital by mid-morning. By 3 p.m., there was still no news. We maintained very cautious optimism, knowing there was a chance that the lungs would not be viable. All of a sudden, the charge nurse burst into my room and said, "They want you in the operating room now!" I clutched her hand and let out every emotion in one big scream: relief, gratitude, fear. The kids were in the quiet room down the hall and heard me scream. My friend explained and Grace fell into her cousin Nathan's arms and burst into tears. They opened their arms and shared the hug with our twins. Grace looked up and said, "It's a miracle."

Having worked as a Child Life Specialist at SickKids and accompanied many patients to the operating room, I was comfortable with what came next. I was not very scared. Quite peaceful, actually. I didn't dwell on the risks. I was ready for this next step. I looked at my doctor, told him I trusted him, and waited while the mask put me into the most important sleep of my life.

It was miraculous. The operation took a little more than eight hours and the surgeon reported that he could see my heart getting stronger as soon as the new lungs were in. It was my first surgical experience and waking up intubated was interesting to say the least! I really wanted the tube out and tried my best to communicate that. Amazingly, the breathing tube was able to come out three days after surgery. I walked in ICU the day after I "woke up"—this was one of the hardest things I've ever done. I kept thinking about the kids with whom I worked at SickKids and said to myself, "if they can do this, then I can do this." It was a powerful motivator.

The transplant program at TGH is a very "well-oiled machine" in terms of the process of care and recovery. I started with 6 chest tubes, and at least 6 lines elsewhere, including 2 in my neck. It seemed that

every day more lines were being removed. I measured my recovery in steps—here is an example:

My sister and friend had been sending out daily emails to family and friends to keep everyone updated on my progress and on this particular day I was able to type the email myself. Here's what I wrote:

***Today's accomplishments include:***

- *typing this email!*
- *1 more chest tube taken out (I now have 3 of 6)*
- *all IVs out (have the big neck one in)*
- *walked to and sat in a chair for an hour. Then walked a little by my bed.*
- *brushed my teeth!*
- *best of all—I am including my first official photo of me without oxygen! (The monkey is hiding the enormous tube in my neck.)*

*Until tomorrow*

*Love Amanda*

I felt very well cared for by the nurses, doctors, and other staff. There was a medication class to learn about the 40-ish pills I take every day. I was oriented to the 3-day-a-week physiotherapy program I was going to attend for the first three months post-transplant. There were weekly clinic visits with bloodwork, x-ray, and pulmonary function testing. In the international community, Canada and TGH are recognized for excellent long-term outcomes because of such close and careful monitoring of lung transplant patients. The hospital staff is a compassionate and skilled multi-disciplinary group of people. That's what I appreciate the most.

I think that the most surreal day came when I was discharged. It seemed absurd to me that after only 2 ½ weeks, I was ready (really ready!!) to go home. Home. A place that I had not spent more than a few hours in since summertime. It was now early November. I walked in without oxygen and climbed our 5 stairs without stopping because of shortness of breath. I walked in a full circle around the kitchen and dining room for the first time. I went down to a part of our basement that I couldn't reach before. I tucked all three kids into their beds at night. All mini-milestones that I treasure and for which I am truly grateful.

As I am writing this article, I am about to reach another milestone. Mid-January will mark 3 months since my transplant. That means finishing physiotherapy and not having another clinic visit for three months! Although I don't have pulmonary hypertension anymore, I feel incredibly connected to the PH community and I am so grateful for the thoughts, prayers, encouraging words, and visits I've received over the last months. Not a day goes by that I don't think about my gift. I have great things planned for these lungs.

*Contributed by: Amanda Littlejohn-English, transplant recipient, Whitby, ON*


*Amanda leaving the hospital!*

*From left to right: 1) Barb, happy event organizer! 2) Participants competing in a Minute-to-Win-It game. 3) Supporters enjoying their evening.*

# Trivia, Fun, Pub and Grub Night: Bringing Community Together over Food, Trivia and Games

I am a person with pulmonary hypertension. I remain pretty stable with the disease and I know how blessed I am. Together with members of the Toronto Chapter, I organized the Trivia, Fun, Pub & Grub Night, which was held on November 7th, 2015 at the Pic-a-Deli Restaurant in Oakville, Ontario. The event included Trivia questions, a few Minute-To-Win-It games, a 50/50 draw and... some great smoked meat sandwiches! This fundraiser has been held the first Saturday night of November for two consecutive years now.

This year, I was the main organizer of the event with wonderful help from Graham and Elsa Beattie; Joan, Jodi and Brooke Paulin; Richard and Patricia Ainsworth; Loretta Chu, our technical wizard; Emily Ho and her husband Stephen; Paul Adams, our MC; and my daughter Meredith. The Beatties put us over the top with ticket sales and prizes, and everyone who participated contributed to making the night a successful event!

My motivation to get involved with organizing the Trivia, Fun, Pub & Grub Night came last year after our Chapter decided not to organize the Vegas Night event that had previously been held as a fundraiser. I wanted to see the Toronto Chapter do a fundraising event for Pulmonary Hypertension Awareness Month and thought it would be a good idea to organize a Trivia Night because I had friends who had been to one and they loved it. I felt a full evening of Trivia on a Saturday night was a little heavy and so included the Minute-To-Win-It games to lighten up the atmosphere. I wanted people to have fun at the event and hopefully come again this year, and it worked!

***I feel that one impact of our event has been to prove that funds can be raised for a cause you believe in and that, if everyone works as a team, it can be a very successful way of spreading the word about PH in your community.***

I had absolutely not a clue how much we'd make the first year and was totally shocked at how much the event raised! This year, we surpassed our goal and raised over $5000... I am extremely happy!

The most memorable moment for me at this year's event was Brooke Paulin telling her story about her experience as a young woman living with pulmonary hypertension. She is such a positive individual and her speech was very moving. People are still commenting how memorable her speech was, and how it gave them a much better understanding of the disease itself and its impact on a patient's life.

I feel that one impact of our event has been to prove that funds can be raised for a cause you believe in, and that, if everyone works as a team, it can be a very successful way of spreading the word about PH in your community. At the 2014 Trivia Night event a man came to the restaurant to purchase his supper and read the sign on the door stating the restaurant was closed for a private function for pulmonary hypertension—lo and behold he'd never heard of pulmonary hypertension until the day before, when he was diagnosed with it! Talk about being where you're meant to be! He has since become a staunch member of our Toronto Chapter! I absolutely would recommend the experience of organizing this kind of fundraiser and would suggest it to other groups as this is a great way of giving back and raising awareness in their area.

*Contributed by: Barbara Heal, PH patient, Burlington, ON*

# Dine for the Cause and Silent Auction: A Great Collaborative Evening

My husband Harry was diagnosed with pulmonary arterial hypertension (PAH) in November 2013. Since then, we have both been involved with PHA Canada: he currently sits on the Board and I am a PHA Canada Ambassador.

This last November, I organized the Third Annual Dine for the Cause and Silent Auction in support of PHA Canada. The event took place on November 18, 2015 at Biagio's Italian Kitchen in Ottawa. This is the third year that a Dine for the Cause fundraiser has been held as a way to bring Ottawa Support Group members, the Ottawa PH Clinic medical team, and supporters together to talk about PH, share information, and have fun. In organizing the event, Carolyn Doyle-Cox was incredibly supportive, and served as primary contact with the PH Clinic team. I am also grateful to Mike Ziola, the owner of Biagio's, his event planning manager, and the restaurant staff, who are a huge help every year.

***It always moves me to tears when I see people come together to support one another and share love with those who are dealing with such a challenging, frightening, and difficult disease.***

This year for the first time, we jointly organized the event with another organization, the Ottawa branch of the Ontario Lung Association (OLA). In addition to being PH Awareness Month, November is Lung Health Month, which made collaborating with the OLA a great fit. The guests that the OLA brought in mingled with the PHA Canada guests and we all learned a bit more about lung health in general, and about PH and PAH in particular. Having dinner together, listening to guest speakers, and hearing the Ottawa Deputy Mayor read the November Awareness Month proclamation was a wonderful way to spend time with others who are also challenged by PH!

My main contact at the OLA was Melanie Estable-Porter, who is an amazing event coordinator and was great to collaborate with! She provided a placement opportunity for a student from the Event Planning program at Algonquin College to help organize a silent auction component. It was the first time I was involved in a silent auction and I was quite nervous about that part! Melanie also arranged for us to be interviewed on Rogers Daytime TV to promote the event!

My goal has always been to use this fundraiser as a relationship and community-building opportunity. I always hope to reach more people than the previous year and, in that sense, we met our goal! The addition of the silent auction component raised money for both charities and was a big success. The PHA Canada team raised $1155 and the OLA a little over $2000.

*Deputy Mayor Mark Taylor and PHA Canada Ambassador Teri Kingston holding the Ottawa PH Awareness Month proclamation certificate.*

Some moments of the evening stand out as memorable. Usually my husband is front and centre with me at the event but this time he could not attend and he provided a message for me to read out to all. I always enjoy hearing the November PH Awareness Month proclamation being read out loud, as well as seeing people learn about PH for the first time. Dr. Lisa Mielniczuk was one of our speakers and she spoke with authority and conviction about new research in the field. However, what was most memorable for me this time was sharing the planning and execution of the event with Melanie, and seeing potential for future collaboration with the OLA. We can learn so much from this organization!

It always moves me to tears when I see people come together to support one another and share love with those who are dealing with such a challenging, frightening, and difficult disease. I love knowing that no one has to face this journey alone. I always say that the PH world is filled with wonderful, amazing, caring people that I wish I had never met. This event was no exception!

*Contributed by: Teri Kingston, PH caregiver and PHA Canada Ambassador, Ottawa, ON*

# PH Tipping Point Stories

Early diagnosis of PH is PHA Canada's number one priority and, through our awareness campaigns, we hope to increase early diagnosis rates. PH is difficult to diagnose because it shares many of its symptoms with other conditions, and we've heard countless accounts of patients being misdiagnosed before receiving an accurate diagnosis of PH. Another challenge to the early diagnosis of PH is the fact that symptoms can develop insidiously, and patients can come to normalize symptoms like breathlessness and fatigue before seeking and receiving a proper diagnosis. We've asked members of our community what is it that brought them over the edge before being diagnosed. Here are some of our members' "PH Tipping Point" stories.

*"**My walk to and from school** was getting more difficult. I thought I was out of shape so I went for a jog. It was that jog (or lack thereof) that made me realize that maybe there was more to it. After a misdiagnosis of asthma, I was finally diagnosed with PH."*

*—Tina Giroulx-Proulx, patient*

*"**First I was told I had asthma** but my legs and ankles were swollen. I was out of breath walking or doing anything for over two years. Then I ended up in the hospital in Calgary."*

*—Sandra Sudo Nelligan, patient*

*"**When, as an infant, her growth was failing** when she should have been thriving."*

*—Stephanie Ricci, mom to Sophia, patient*

*"**When what should have been a 45-minute bike ride** was now taking two hours."*

*—Derek Henderson, patient*

*"**I was 27 years old when I was diagnosed,** despite being born with a hypoplastic right lung. I got an X-ray from a clinic in a town of 200 when I went to their emergency because I had severe pneumonia. Health care in my area is not great..."*

*—Jodie Ashini, patient*

*"One day stands out as a turning point for me. I got up from my bed and I could barely breathe trying to walk across the hallway from my room."*

*—Joan Nemeth, patient*

*"One day while running around, Riley started to show signs of distress. He was having trouble breathing and his lips turned blue."*

*—Danush Rudolph, mom to Riley, patient*

*"When I came in from a night out for my birth-day, and was so short of breath that I couldn't even finish my dinner. My feet were swollen like I had never seen them before."*

*—Loretta Chu, patient*

*"When I was coughing so much that I was not able to take more than 1 call during my entire call centre shift."*

*—Adam Raeburn, patient*

*"When my shortness of breath peaked and I could no longer walk even 5 feet without panting and stopping to catch my breath."*

*—Nicole Dempsey, patient*

*"I could no longer climb the stairs to my apartment. I could not pick up my kids' toys on the floor without getting very dizzy. In one year, my heart had doubled on an X-ray."*

*—Sandy Vachon, patient*

*"I thought that I was just taking a long time to get over a case of pneumonia. Then, I ended up in emergency with chest pain and shortness of breath."*

*—Jas James, patient*

*"When my heart began to feel like a sack of gerbils in my chest and my fingertips turned blue."*

*—Jill Morton, patient*

# Chronic Thromboembolic Pulmonary Hypertension: Curable Type of Pulmonary Hypertension

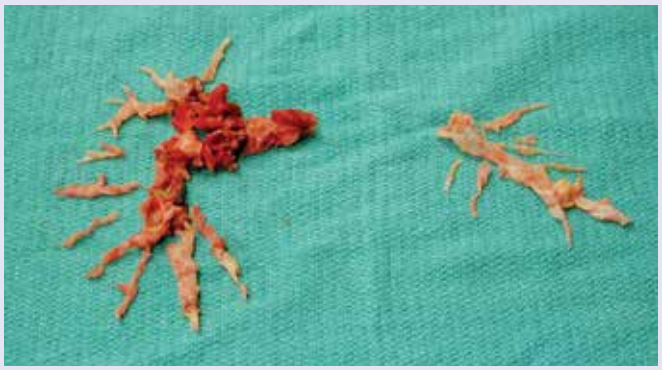
***Anastasia Bykova is the Nurse Practitioner at the Toronto CTEPH Program, Division of Thoracic Surgery, Toronto General Hospital, who follows patients diagnosed with CTEPH before and after their pulmonary endarterectomy surgery. Besides her clinical practice, Anastasia is actively involved in patient and health professional education about CTEPH, raising awareness of the diagnosis, and academic work on this topic.***

Chronic thromboembolic pulmonary hypertension (CTEPH) is a type of pulmonary hypertension (PH) that is caused by chronic blood clots inside the arteries of the lungs. CTEPH is the only type of PH that is curable by a surgical procedure called pulmonary endarterectomy (PEA). The complex disease process in CTEPH poses a challenge for a newly diagnosed patient and their family to understand what is happening inside the patient's body. Therefore, I would like to talk about how CTEPH develops and how it is treated.

## DIFFERENCE BETWEEN ACUTE AND CHRONIC PULMONARY EMBOLI

When a blood clot forms inside a deep vein of the legs, it can break off, travelling to the heart and settling inside the arteries of the lungs. This process is called **acute pulmonary emboli (acute PE)** (see image *Acute Pulmonary Emboli*). Our bodies have the natural ability to dissolve blood clots, and people who are diagnosed with PE will be treated with anticoagulation medications or blood thinners to help their bodies get rid of these clots. The bodies of most patients are able to dissolve pulmonary emboli by the end of treatment with anticoagulation medication, with no long-term consequences to their health. However, in 3-5% of cases, the blood clots do not dissolve despite the anticoagulation treatment and create fibrous scar tissue inside the pulmonary arteries. This process is called **chronic pulmonary emboli** (see image *Chronic Pulmonary Emboli*). It is still unknown why some people are able to dissolve blood clots while others are not.

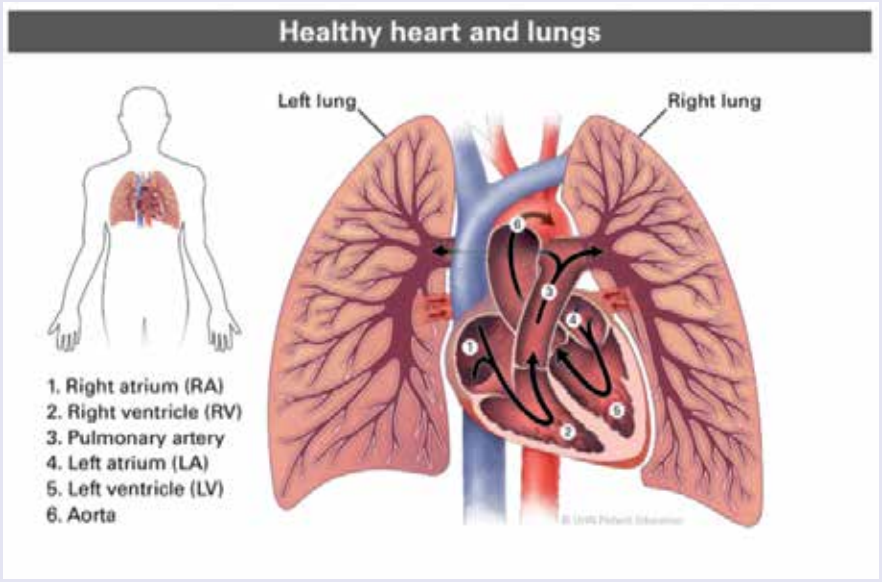
***Acute Pulmonary Emboli (Acute PE)***
*Fresh blood clot tissue removed from the pulmonary arteries during the surgery called thrombectomy.*
*Image used with permission from the Toronto CTEPH Program.*

***Chronic Pulmonary Emboli***
*Fresh blood clot transforms into fibrous scar tissue that lines the pulmonary arteries in CTEPH. Chronic pulmonary emboli removed from right and left lungs during the surgery called pulmonary endarterectomy.*
*Image used with permission from the Toronto CTEPH Program.*

## HEALTHY HEART AND LUNGS

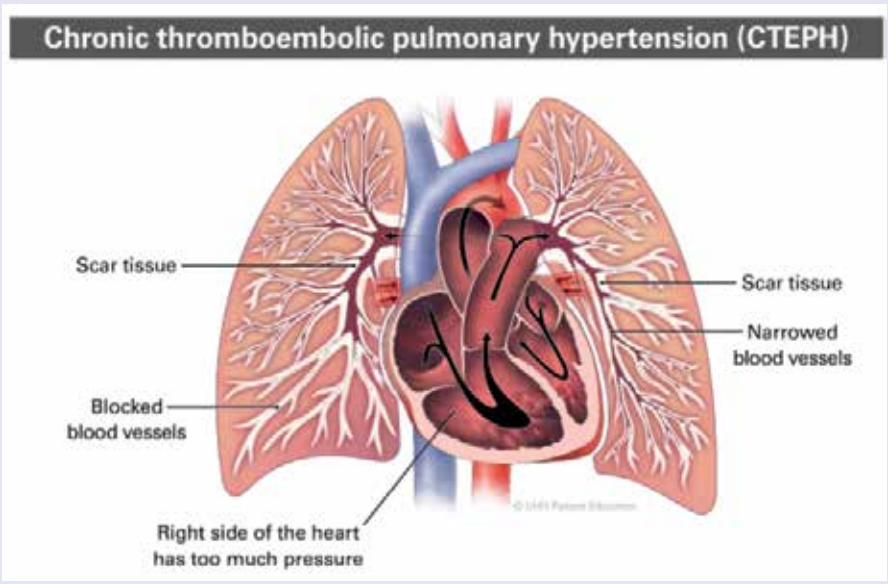
In order to understand what happens to the body in CTEPH, let's look first at the anatomy and function of healthy heart and lungs. The blood rich in carbon dioxide travels from the entire body, including arms, legs, and head, to return to the right atrium of the heart (see image *Health Heart and Lungs*). From the right atrium, the blood travels to the right ventricle and fills it up until the right ventricle contracts and pumps the blood into the pulmonary arteries. While travelling in the pulmonary arteries, the blood gives away carbon dioxide and receives oxygen. Oxygen-rich blood then returns to the heart through the left atrium and fills up the left ventricle. When the left ventricle is full and contracts, the oxygen-rich blood is pumped out to the aorta and to the rest of the body.




***Healthy Heart and Lungs***
*Image used with permission from University Health Network Patient and Family Education Program.*
*© University Health Network, Patient & Family Education.*

## HEART AND LUNGS IN CTEPH

In the lungs affected by CTEPH, chronic blood clots inside the pulmonary arteries create scar tissue leading to the narrowing of the pulmonary arteries and physical obstruction of the blood flow (see image *Chronic Thromboembolic Pulmonary Hypertension*). As a result, the pressure in the lungs increases, causing PH. With the elevated pressure inside the pulmonary arteries, the right ventricle must generate more force to pump blood against blocked blood vessels. Over time, the right ventricle suffers from the high pressure it has to generate to push blood forward, which leads to thicker walls and a dilated chamber. This is called **right heart failure**. As the right side of the heart struggles to push blood forward, the fluid accumulates in the right ventricle and atrium, leading to back flow into the other parts of the body such as legs and abdomen.




***Chronic Thromboembolic Pulmonary Hypertension (CTEPH)***
*Image used with permission from University Health Network Patient and Family Education Program.*
*© University Health Network, Patient & Family Education.*

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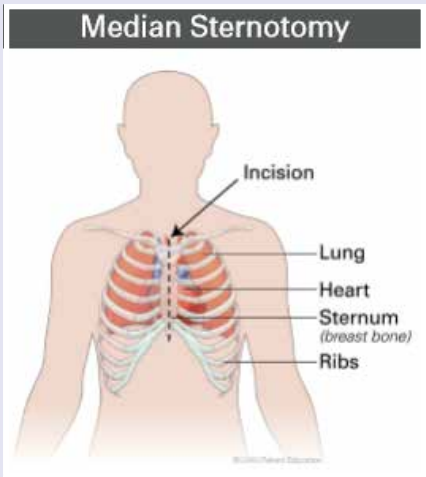
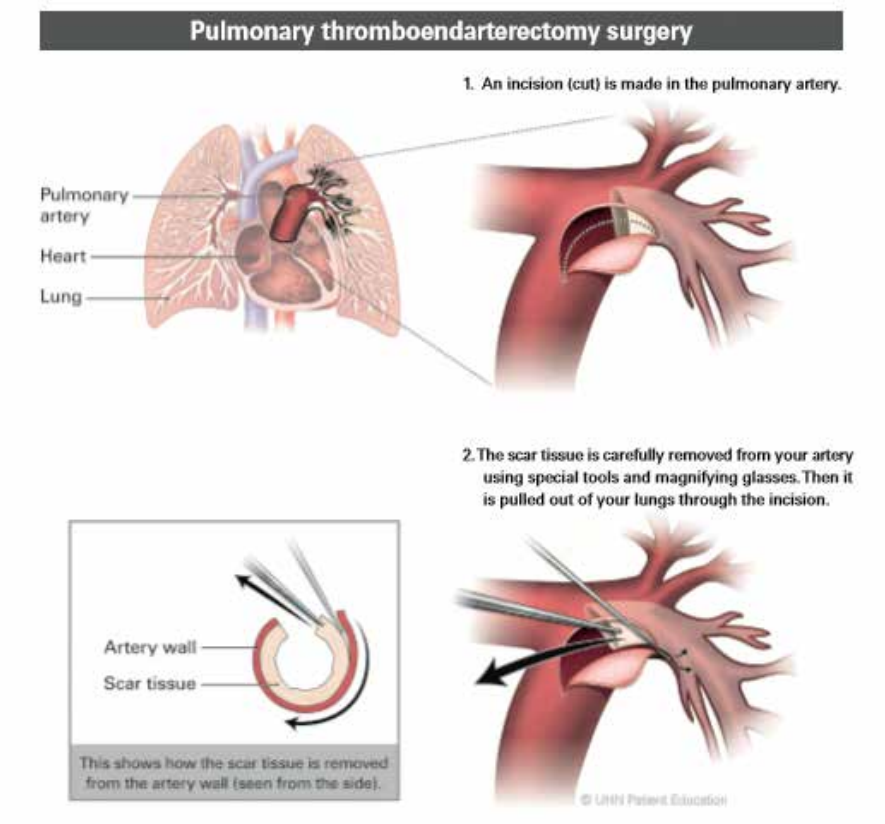



***Median Sternotomy***
*Image used with permission from University Health Network Patient and Family Education Program.*
*© University Health Network, Patient & Family Education.*

Initially, the majority of patients with CTEPH have non-specific symptoms of shortness of breath upon exertion, and fatigue from the chronic blood clots and PH. As the disease progresses, patients find that shortness of breath becomes worse, limiting their daily activities (i.e. exercising, grocery shopping, cleaning the house, mowing the lawn). Other symptoms of advanced disease are syncope, chest pain, and swelling of the legs and abdomen. Without treatment, the survival rate of a patient with CTEPH is poor.

## TREATMENTS FOR CTEPH

CTEPH is unique in the way that it is the only type of PH that is potentially curable by a surgery called **pulmonary endarterectomy (PEA)**. During PEA surgery, a surgeon makes a cut through the sternum (breast bone) to expose the heart, major blood vessels including the pulmonary artery, and parts of the lungs (see *Median Sternotomy* image). The patient goes onto the heart-lung machine and is cooled down to 18-20°C to slow down the metabolism of vital organs. The surgeon then makes a cut in the pulmonary artery to open it up (see *PEA Surgery* image). The fibrous scar tissue is located inside the pulmonary arteries and is carefully removed by the surgeon using special instruments and magnifying glasses. When all chronic clots that the surgeon can find are removed, the cut in the artery wall is closed together with sutures. The goals of PEA surgery are: to cure PH; to treat shortness of breath and improve quality of life so that the patient can return to a physically active lifestyle; and to prevent right heart failure. Most patients who wake up after the surgery report that their breathing is much better and that their shortness of breath has gone away.




***Pulmonary Thomboendarterectomy Surgery***
*Image used with permission from University Health Network Patient and Family Education Program.*
*© University Health Network, Patient & Family Education.*

Medical therapy with PH-specific medications is sometimes used to treat a select group of patients with CTEPH. Riociguat® is the only medication that has been approved by Health Canada for the treatment of CTEPH when patients are not surgical candidates for PEA or for patients with residual/recurrent PH following PEA surgery. The goal of medical therapy is to slow down the disease progression.

Patients who are diagnosed with CTEPH often have significant physical limitations due to shortness of breath, causing poor quality of life. Without surgery, CTEPH is life-threatening. In the hands of an experienced CTEPH team, most patients following PEA surgery are cured of their disease, and are able to breathe normally and enjoy an active lifestyle.

*Contributed by: Anastasia Bykova, BScN, MN, RN (EC), Nurse Practitioner, Division of Thoracic Surgery , University Health Network, Toronto, ON.*

# Advocacy Focus: Why Your Voice Matters

In his "Message from the Chair," Dr. Sanjay Mehta states that one of PHA Canada's top priorities is ensuring that all PH patients in Canada have access to the treatment that is right for them. As an organization that strives to achieve a better life for all Canadians affected by PH, it is critical that we advocate on behalf of patients and provide tools that empower the PH community to make its voice heard. We believe that all patients—no matter their financial situation, province of residence, or private insurance coverage—should have access to the medications that their PH specialist thinks will provide the best possible outcomes.

In recent months, we have devoted resources and energy to voicing our concerns and asking provincial governments to address the challenges to treatment accessibility that PAH patients in Canada (with the exception of Quebec residents) currently face. Because PAH is a very complex, serious, progressive, and potentially fatal disease, we believe that it is of the outmost importance that treatment decisions remain in the hands of PAH experts, as they are best positioned to assess which medications—amongst all those approved by Health Canada for the treatment of PAH—will provide optimal treatment for each unique patient.

However, recent recommendations published by the Canadian Agency for Drugs and Technologies in Health (CADTH) favour a stepped approach to initial PAH therapy, which may impact newly diagnosed patients' access to optimal treatment. Further, as explained by Dr. Mehta, the closure of negotiations between the pan-Canadian Pharmaceutical Alliance (pCPA) and the manufacturer of Opsumit (macitentan) in December 2015 means that this treatment, though approved by Health Canada in 2013, is currently still not available as a treatment option for PAH patients (outside of Quebec) who rely on public funding.

Since June 2015, PHA Canada—with the invaluable help of our members, Ambassadors, support group leaders, and supporters—has been asking provincial governments across the country to disregard CADTH's recommendations and ensure that newly diagnosed patients have access to the therapies their PH specialist feels will be most effective. As part of our ongoing efforts to ensure that PH patients have access to optimal treatment, we have also brought attention to the lack of access to Opsumit faced by a large portion of PAH patients in Canada.

Many of you have contributed to our advocacy efforts: supporters across Canada have so far sent over 1,000 letters to their provincial governments; our Ambassadors and members are meeting with their provincial representatives to discuss issues regarding treatment accessibility; and our members have been featured in multiple media pieces that create greater awareness of the challenges the PH community faces. PHA Canada has also partnered with the Scleroderma Society of Canada (SSC) in order to increase the reach of our advocacy campaign, including co-hosting a PAH Advocacy Day at the Ontario Legislature on December 03, 2015.

In the following pages, Nicole Dempsey, Ruth Dolan, and Joan Paulin share how empowering it has been, since being introduced to PH, to become strong advocates for the PH community. Their stories are preceded by an informative interview with Dr. Mehta, initiated by PAH patient Serena Lawrence in an effort to better understand the challenges to treatment accessibility in Canada. PAH patient Brooke Paulin's story completes the Advocacy Focus segment by highlighting the importance of making your voice heard as part of our community's fight for treatment accessibility.

We hope that these articles will inspire you to lend your voice—whether again or for the first time—to our advocacy efforts. Now is the time to roll up our collective sleeves and show just how strong and united the Canadian PH community is! Together with the SSC, we have developed tools to help our members and supporters make their voices heard. Please visit **www.takeactionPAH.ca** to learn more about our current advocacy campaign and how you can get involved. This includes: a new email tool that enables you to easily ask your provincial government to address the needs of PAH patients; resources for those who are ready to take the next step and meet with their provincial representative; and tips for anyone who wants to share their story with the media. We're also always happy to provide individualized support to anyone who wants to be involved. We believe that patients should come PHirst, and we need your help communicating that to decision makers!

**www.takeactionPAH.ca**

*Contributed by: Mariane Bourcheix-Laporte, PHA Canada Communications Associate*

*Advocates participating in PAH Action Day, December 3rd, 2015.*

# Accessibility of PAH Therapies in Canada Part I

INTERVIEW WITH DR. SANJAY MEHTA BY SERENA LAWRENCE, PH PATIENT

***In March 2015, the Canadian Agency for Drugs and Technologies in Health (CADTH) suggested that all newly diagnosed people with Pulmonary Arterial Hypertension (PAH) in functional class II or III receive the same mono-therapy approach for their treatment. This approach would take the decisions for treatment out of the hands of PH experts and place it in the hands of government bureaucrats. I asked PH specialist Dr. Sanjay Mehta to answer some questions I have regarding the accessibility of PH therapies in Canada.***

## *ABOUT THE CONTRIBUTORS:*

***Dr. Sanjay Mehta, MD, FRCPC, FCCP*** *(left), is Professor of Medicine at the University of Western Ontario, and Director of the Southwest Ontario Pulmonary Hypertension Clinic at the London Health Sciences Center in London, Canada. He also serves as the PHA Canada Chair.*

***Serena Lawrence, Kitchener, Ontario*** *(right), was diagnosed with PAH in 2013. She is a freelance writer who runs the PHight or Flight Project, a blog that shares the exceptional stories of other people with PH. She also blogs about her experience as a young adult with PH. Please visit: phightorflight.blogspot.ca.*

***Serena:*** *I know that you are very involved with and passionate about the PH community, and that you have been working very hard to ensure that all newly diagnosed patients have access to the therapy that is best suited to their needs.*

***Could you briefly explain why it is so important that PH specialists, like yourself, are able to treat PH patients on an individual basis and recommend a therapy suited to their needs instead of the stepwise and rigid therapy approach suggested by CADTH?***

***Dr. Mehta:*** It is one of the most basic principles of medicine: that each patient is an individual, and needs to be understood, respected, and treated as an individual. This includes doctors making decisions about the best medical treatment for a patient's illness. Although many patients may have the same illness, as in the case of PH, they are all still unique individuals. As such, there is no reason to expect that they respond similarly to PH medications, or that the same medication is the best one for each patient. Expert PH physicians need to consider many factors in deciding on the best initial and subsequent medical treatment for each PH patient. The decision is based on having expertise and experience in PH, understanding the patient and their other conditions, and the specific risks of each treatment. Even in clinical studies of PH medications, all patient participants don't respond similarly, and while some may respond well, others may not. Importantly, in dealing with a serious, progressive illness like PH, a patient can't afford to waste precious time "trying" anything other than the most effective, best treatment for them.

***Serena:*** *One of the newest Health Canada-approved oral medication for PAH, macitentan (Opsumit) is not available to most Canadian PAH patients, as per the recent pan-Canadian Pharmaceutical Alliance (pCPA) decision to close negotiations with the manufacturer, with the impact that this drug is not approved for funding in most of Canada.*

***I understand that Opsumit is generally well tolerated, and that it was tested in one of the largest and longest clinical studies of any approved PH treatment. Can you discuss some of the benefits of Opsumit? If the government continues to deny funding for Opsumit, what is the future for this medication in Canada?***

***Dr. Mehta:*** In 2013, the SERAPHIN study reported on the benefits of macitentan (Opsumit) in what was at the time the largest, longest study ever in PAH patients. It showed that macitentan reduced the morbidity (severity of illness) in PAH patients, specifically reducing the risk of progressive worsening of PAH by 45%, and reducing the risk of hospitalization by 50% over 3 years. Moreover, treatment with macitentan significantly improved symptoms, quality of life, and exercise capacity in PAH patients. This is a unique study that showed for the first time the long-term benefits of treatment with a PAH medication, compared to all other PAH studies, which only looked at benefits over 3-6 months. Clearly, PAH patients don't want to just improve over the short-term, but hopefully remain well for many years!

As a result, treatment with macitentan has been strongly recommended by the most recent 2014 PH Clinical Practice Guidelines, jointly published by the European Society of Cardiology and the European Respiratory Society. Moreover, macitentan has been approved for funding in the US, and in many European countries, and many PAH patients are currently being treated with macitentan. Notably in Canada, Quebec approved public funding of macitentan in October of 2013, such that Quebec PAH patients have complete access to macitentan as a treatment for their PAH, should their expert PH physician decide it is the best treatment for them. Similarly, several large private insurance companies across Canada have approved macitentan coverage for their clients. However, pCPA recently denied funding for macitentan for the rest of Canadians (PAH patients living in all the other provinces and territories who do not have access to private health care), and indeed, has broken off negotiations with the pharmaceutical manufacturer, Actelion.

This is a very concerning development for Canadian PAH patients, most of whom have been denied access to public funding for treatment with macitentan. Moreover, this establishes a dangerous precedent whereby any and all future new PAH therapies (for example, the new oral selexipag or Uptravi, which was just approved by the FDA in the US and by Health Canada) may similarly not get approval for funding for Canadian PAH patients. It would appear that Canadian and provincial governments are saying that PAH patients are doing just fine with the therapies they have available today! Clearly, PAH patients and their physicians know otherwise; despite treatment with the many PAH medications we have, many PAH patients remain seriously ill, limited in everyday life, and their disease continues to progress until it takes their lives, on average 7-10 years after diagnosis. Is it reasonable to accept that? Should we not try to further improve the health and lives of PAH patients? Most definitely, all Canadian PAH patients and their caregivers would want us to continue to develop, test, approve, and make available newer and better PAH treatments. Lack of government understanding of this critical issue is already leading to less than optimal treatment of Canadian PAH patients, as currently demonstrated by lack of access to macitentan, and is likely to reoccur with lack of availability of future new PAH treatments.

***Serena:*** *Thank you for sharing your thoughts on this complex issue. Is there anything that you would like to share going forward?*

***Dr. Mehta:*** All Canadian PAH patients and their caregivers should be heartened by the incredible progress we've made in the treatment of PAH, since the 1st medication, intravenous epoprostenol (Flolan), became available in 1997. Currently, 9 different medications are approved and generally available for the treatment of PAH in Canada. As a result, the quality of life and survival of most PAH patients have significantly improved. This is important to keep in mind and we should remain hopeful that, thanks to the joint efforts of dedicated advocates, members of the medical community, and PHA Canada, new treatments such as Opsumit will be made available to all PH patients in Canada. This is what we must continue to strive for.

*Fierce advocate and PHA Canada Ambassador Nicole Dempsey meeting with her MPP Kathryn McGarry in the summer of 2015.*

# Giving a Face to PH

I was diagnosed with IPAH in December 2013 and became an Ambassador for PHA Canada in August of 2014. Since my diagnosis, I have been very fortunate in the fact that I was prescribed two PH medications (one oral and one IV) that have been both working at keeping me stable. I became involved in PHA Canada's advocacy efforts because I want everyone in the PH community to have this opportunity as well. If newer treatments, like Opsumit, aren't accessible to patients, this could mean denying a patient life-saving medication. I think it's absolutely imperative that treatment decisions be left in the hands of PH specialists, and that's the message I and other advocates have been sending to provincial governments.

I was very excited to take part in the PAH Action Day. I went to Queen's Park not quite knowing what to expect, and it was certainly an amazing experience. I knew I was going to be speaking along with Dr. Mehta, Ruth Dolan and Anna McCusker from the Scleroderma Society of Canada, but I didn't quite know how the media press conference worked. It was very empowering in some way to tell my story and to express the importance of the reason we were there: to ensure that all Health Canada-approved PAH therapies be accessible to PAH patients at diagnosis and beyond.

After the press conference, I was interviewed by EPOCH times and the Globe and Mail. I then had the opportunity to meet with my MPP, Kathryn McGarry, whom I had met at her local office in the summer to discuss the need for access to all Health Canada-approved PAH medications. At Queen's Park, I also met with MPP and Associate Health Critic for Long-Term Health Care, Bill Walker. I thought that the MPPs seemed genuinely interested in helping and providing action for PAH patients.

***It was very empowering in some way to tell my story and to express the importance of the reason we were there: to ensure that all Health Canada-approved PAH therapies be accessible to PAH patients at diagnosis and beyond.***

As an Ambassador for PHA Canada, I've had a few opportunities to advocate. I've learned that advocating isn't difficult. It can be as simple as being present with other members, providing your voice, and telling your story. As a patient, I think it's important to give a face to PH and provide a voice for this rare disease. It felt good to be at Queen's Park and speak on behalf of the PH community.

*Contributed by Nicole Dempsey, PH patient and PHA Canada Ambassador, Cambridge ON*

*Ruth (centre) presenting at the PAH Advocacy Day press conference.*

# PHighting for Those Affected by PH: What I've Learned on my Path to Advocacy

I became aware of PH in 2007 when my youngest daughter was diagnosed with it. I joined PHA Canada's Toronto Chapter Support Group in 2008, and was invited to become the co-leader of that group in the spring of 2010. I now sit on PHA Canada's Board, serve as Liaison with Ambassadors, and am involved in organizing fund and awareness raising activities.

In 2008-2009, when I first became more involved with the Toronto Chapter, the PH community in Ontario was fighting to ensure that combination therapy would continue to be covered by the Ontario Public Drug Plan. This experience made me aware of how important it is to continually monitor—and be ready to challenge—provincial governments when they make decisions that can threaten patients' lives, i.e. to discontinue funding for, or to not fund, treatments that are needed by PH patients.

Regarding the current issues we face with ensuring access to treatment, I feel one thing we need to do is focus our efforts on creating much greater awareness of PH and the issue of access in particular. Ambassadors, Support Group leaders, and the members of our community need to be encouraged, inspired, and energized to reach out to more health care providers, government representatives, and media outlets in this regard. When I am troubled and discouraged by the minimal response we get for a call to action, the question in my mind is: "What would it mean to you (PH patient/caregiver) if you could not get the treatments you/your loved one need/s?" To our community I ask: "What would you be willing to do to help ensure that new and better treatments continue to be available?" I also ask myself: "Who is going to stand up for us if not members of the PH community!?"

***"Doing" awareness and advocacy IS empowering! I feel much less helpless than I once did when our daughter was first diagnosed. I am more knowledgeable about PH and what those affected by PH need for a better quality of life.***

## PAH ADVOCACY DAY AT QUEEN'S PARK

My involvement with the Ontario Lung Association had previously brought me to speak to elected officials at the Ontario Legislature so I felt comfortable participating in PAH Advocacy Day at Queen's Park.

It's not as intimidating as it looks, especially when this type of action is well supported and when participants are well trained. Before the day's events, I felt excited for the opportunity to speak about the specific access to Opsumit issue. As the day unfolded, I felt very pleased with how well it was organized and supported—that made me feel reasonably confident that I could do my best possible job! I think I can speak for everyone that was present and say that it was a great learning experience that increased our self-confidence to take on more advocacy outreach! After the media conference, I had the opportunity to meet with NDP Health Critic France Gélinas, who seemed very receptive to our "ask." Earlier in the week, I had met three other MPPs during Lung Health Day, which was organized by the OLA. I did not speak to any media at Queen's Park, but my local paper, the Bradford Times, published an article online after the event.

## TIPS AND TRICKS FOR EFFECTIVE ADVOCACY

Over the years and the meetings I have had with my, as well as other MPPs, I have learned to focus on the "asks"—the main reasons for the meeting. I also always tell my PH Story because it helps the person I'm speaking to relate to what I have to say. Thanks to the support and training I have been given through PHA Canada, it has been much easier than I would have imagined. And with practice it gets easier! I have developed the confidence to do more networking and to explore other opportunities to get the word out about PH and fight for our community.

I encourage everyone who has been affected by PH to get involved! Your voice matters and you can start speaking up about the needs of PH patients and caregivers (including access to treatment and treatment innovation) in any way you can. Advocating can be as easy as sending a letter to your Health Minister, or more involved like reaching out to health care providers or meeting with your elected Member of Parliament. Here are a few tips if you do meet your provincial representative: make sure to bring someone who can be there to support you, take notes, and help you "debrief" after the meeting; write a report on your experience and share it with PHA Canada and other PHighters; write to your local paper about your visit; take a photo with you MPP and share it on social media; and always send a thank you note or email following a visit.

## MAKING A DIFFERENCE

What I find most rewarding about my advocacy experience is feeling that I might be making a difference in the lives of PH patients and their caregivers. I believe I am empowered through all of the activities in which I participate (awareness, advocacy, education, and patient support). Over the years, I have made so many connections and new PHriends that I would not have made had I not been so involved. "Doing" awareness and advocacy IS empowering! I feel much less helpless than I once did when our daughter was first diagnosed. I am more knowledgeable about PH and what those affected by PH need for a better quality of life. I want to share that knowledge with others, especially those who can make a difference: MPPs, health care providers, the Minister of Health, and even the Premier. I only wish that we could find a way to inspire many more to join us in this PHight!

*Contributed by: Ruth Dolan, PH caregiver and PHA Canada Board member, Bradford, ON*

*Ruth, photographed at the PAH Advocacy Day with NDP Health Critic & Chair of the Lung Health Caucus France Gélinas (far left), Scleroderma Society Executive Director Anna McCuster (second to left), and PAH specialist Dr. John Granton.*

*Unstoppable advocate Ruth presenting at the closing ceremony of the Bradford Run/Walk, Swim or Bike/Spin PH Challenge in November 2015.*

# From Ice-Skating Athlete to PH Advocate: My Journey as a Young PH Patient

*Brooke (far right) with her mother Joan (centre) at the GolPH for PH fundraiser held in July 2015.*

I was diagnosed with PAH in March 2014 at the age of 24. My life has completely changed since my diagnosis. Even though I don't look sick, I am unable to do things young adults take for granted. I can no longer do any strenuous exercise—that means no running, no biking, no weight-lifting to keep my muscles strong. Before I was placed on life-saving medication, I could barely make it up a flight of stairs or walk from my car in the parking lot to my desk at work. I am currently eligible to have a handicapped sticker for my car. Of the many things I have become since diagnosis, one is more tolerant of those whom I used to think were taking advantage of handicapped parking when they had no visible handicap—and now that could be me!

Up until I was 21 years old, I was a high-performing athlete. I spent my entire childhood on the ice—4, 6, 8 hours a day. By age 14, I was a national level athlete in both singles and pairs figure skating. I represented Canada at several international skating events. I spent hours in the gym when I wasn't on the ice. My aerobic capacity was second to none.

I rationalized that the reason I could not get up a flight of stairs was because I was no longer a competitive athlete. I had never been "out of shape." This had to be what "out of shape" meant. I started going back to the gym in late 2013, but I struggled with even the "easy" things like walking on the treadmill for more than a few minutes. But it wasn't until I was walking on a street in downtown Toronto in February 2014, on a cold winter's day, that I knew something was wrong. I had to stop walking because I couldn't catch my breath. I literally struggled just to breathe. I went to the ER that day, thinking I might have asthma, and did not leave the hospital for 10 days. This was not asthma!

While I don't look any different on the outside than I did when I was an athlete, I am living a new "normal." I still struggle with my new normal. It comes in waves: good health days, not so good health days, good emotional weeks, not so good emotional weeks. I am learning a lot about myself. I am learning to live one day at a time, to not think of the things that I will not be able to do or have—children, for example. And I am learning to be very thankful. Thankful for family and true friends; thankful for great doctors and terrific research into PH; thankful for a health care system that ensures I have access to expensive medications that I cannot afford to pay for; thankful to live near Toronto where some of the best respirologists and cardiologists practice; thankful for a support group that meets every month and reminds me that, despite having PH, we can still laugh. And I am mostly thankful for the medications that are currently available in Canada to treat PH. There are currently nine approved treatments that target three known pathways in which PH affects the lungs. Some come in pill-form and others are extremely invasive. None of them cure PH and not all of them work for every PH patient. But if your doctor can find the right treatment for you, it can slow down the progression of the disease. And that's where everyone can get involved in the cause. PH is still a rare disease with few advocates (unlike a disease like cancer) and with challenges to treatment accessibility, the PH community needs all the support it can get.

***I am learning to be very thankful. Thankful for family and true friends; thankful for great doctors and terrific research into PH; thankful for a healthcare system that ensures I have access to expensive medications that I cannot afford to pay for; thankful to live near Toronto where some of the best respirologists and cardiologists practice; thankful for a support group that meets every month and reminds me that, despite having PH, we can still laugh.***

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Currently, my PH doctor has the ability to determine, based on my symptoms, which of the nine medications he thinks, in his professional judgement, would work best for me. I am so thankful for that because I am currently on two medications that target two of the possible three paths. I felt immediate improvement after I was placed on the first medication. Had it been left there, I would never have known that I could feel even better. But I do and it is because my doctor put me on a second medicine. I have been on these medicines since my diagnosis and they have saved my life. In my support group of eleven or twelve people, not one of us is on the same combination of medicines—but each combination is "saving" a member's life.

As an aside, this says something about how complicated PH is, and how difficult it will be to find a cure. PH doctors know that the key to long-term success with this disease is to hit it hard initially because disease progression cannot be reversed.

In a bid to reduce health care costs, the Ontario government (as well as other provincial governments outside of Quebec) is looking at forcing PH doctors to adopt a "one-medicine-fits-all" approach to treatment for newly diagnosed patients. If I was being initially diagnosed in this new world and did not have private insurance, I would have access to only certain medications, and only when my health began to decline again would my doctor be allowed to put me on other medications. Even if I currently have access to effective treatment for me, because of the fact that not all PH treatments are accessible to all PH patients in Canada, I am concerned that, if and when my PH doctor has to change my medication (because my disease

has either progressed or my medications are no longer working effectively) I will not be given the "best" available medication, but the cheapest next alternative.

PHA Canada is asking the Ontario government (and other provincial governments) to rethink this strategy but we are a small group with a small voice. Anyone that has been touched by the struggles of the PH community can lend their voice to our cause by sending an email to their Premier and Minister of Health using the online tool found at www.takeactionPAH.ca. This tool makes it simple to ask the government to allow doctors, who are the people with the medical expertise, to provide individualized treatment to each patient and not adopt a standard one-medicine-fits-all approach.

There is another way people can help: by signing a donor card or going online to register as an organ donor. PH is a disease that affects the lungs and the heart. When medicines no longer work to stop the progression of the disease, PH patients who are healthy enough are considered for both heart and lung transplants. There are way too few organ donors and not all patients on transplant lists get the life-saving call they are waiting for.

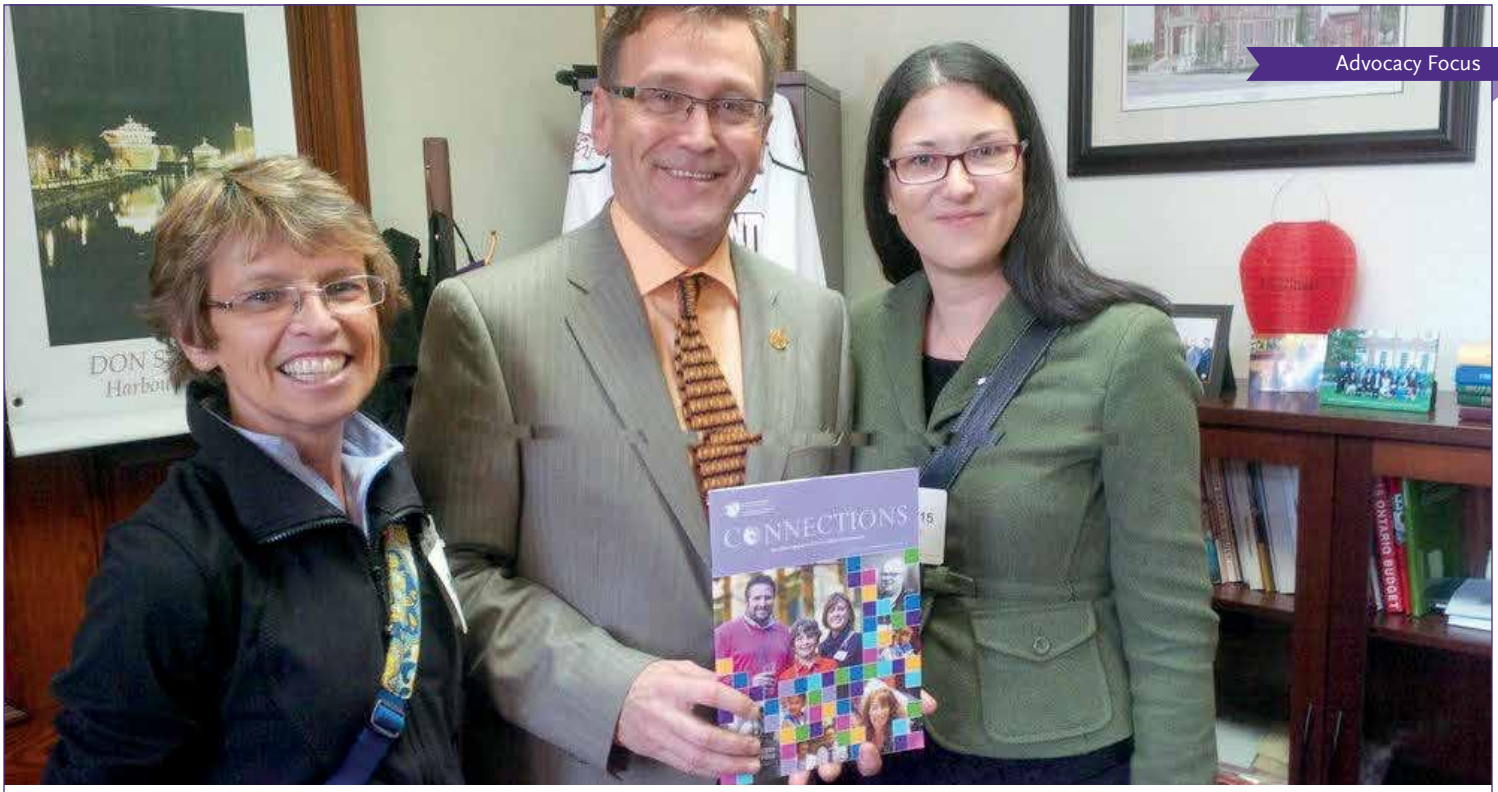
In my PH journey, amongst the many wonderful people I have met and learned so much from, I draw strength from an inspirational lady so full of life: a Mom with two young children who was a teacher until her health forced her to the sidelines. She recently underwent a double lung transplant. After a long journey in the hospital, she returned home to her family and is adapting well to her new lungs and new life. I can guarantee you that she is so thankful to be alive and to get a second chance at life. I don't know if any of my organs will be transplantable, but I have signed my donor card.

In closing, I would like to say that I see life through a different lens now. Sometimes that hurts, but it does bring clarity that most young people don't have. And, yes, I am thankful for that too.

*Contributed by: Brooke Paulin, PH patient, Mississauga, ON.*

*Text adapted from a speech given at the Trivia Fun, Pub, and Grub Night, November 7, 2015.*

*Brooke spinning in the air at an ice skating practice.*




*Joan with MPP Bill Walker and PHA Canada advocate Nicole Dempsey at the Queen's Park PAH Advocacy Day, December 3rd, 2015.*

# My Journey as PH Caregiver and Advocate

My daughter, Brooke, was diagnosed with severe PAH at the age of 24, in March 2014. It would be an understatement to say that we were all devastated. It felt like the floor had dropped out of our lives. Brooke had been a competitive national-level athlete until she was 21. How does someone with cardiac capacity second to none become someone who can barely climb a flight of stairs within three years? We still don't know how and probably never will. But we do realize now that the signs had been there for at least a year—she struggled riding in a charity bike ride in the spring of 2013, and playing volleyball in a charity tournament that summer; she felt out of breath volunteering at a skating event in January 2014; and finally, she literally could not catch her breath on a walk to a bar to watch the men's Olympic gold medal hockey game in February 2014. That night she was admitted to the hospital for a ten-day stay on the cardiac floor. That's where we first heard the words "pulmonary hypertension."

The days and weeks that followed are a haze. There were no "good" days for weeks, maybe months. It's hard to see good when your daughter's life seems to be crashing around her. She was struggling with the diagnosis; I was struggling with both the diagnosis and the need to "keep a stiff upper lip." She needed a shoulder to cry on; a rock to hold on to. I had to be that shoulder, that rock. Would there ever be a day where my first waking thought wouldn't be about Brooke and PH? I couldn't see it then.

Like most people, after we first heard the words pulmonary hypertension, we turned to the Internet. We read many things—most of them scary—, which only made our states of mind worse. But we did find some useful information. We found PHA Canada, PHA in the United States, and several Facebook pages (both here in Canada and in the US)—one of which was a support group for PH patients and caregivers in the Toronto area. We immediately became members of both PHA Canada and PHA in the US. We get valuable and up-to-date information from both sources. We also go to Toronto Support Group meetings every month and appreciate the efforts of Ruth Dolan and Loretta Chu to keep us all informed. Because PH is a rare disease, it is good to be able to talk to people about what is happening in the PH community. We know we are lucky to live in a larger community close to Toronto.

It took a long time but I no longer wake up every morning with PH on my mind. Brooke's Dad and I wear our "Phenomenal Hope" bracelets 24/7. Brooke, her Dad, and I went to the PH Conference in Montreal in May. I have to be honest, Brooke didn't really want to go as she wasn't sure she was ready to see "what lay ahead for her." But it was an amazing experience for all of us. We saw and met people "living" with PH. There was no sadness. The speeches were uplifting and the sessions were informative. Brooke was so glad she went. We were all glad we went—so much so that we are hoping to go to the US Conference this year.

We have, over time, gotten more involved in PH advocacy. There are so few of us and we need to make sure that our voices are heard. Brooke's Dad organized a golf tournament last summer, Golph for PH, which was very successful. It was held at a golf course in Mississauga. The second annual Golph for PH will be held this July. If you live in the area and you golf, we'd love for you to join us! For the past two Novembers, I have helped run our Trivia, Fun, Pub, and Grub

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Night in Oakville. If you live in the area and are looking for a fun night out, come join us next November. We get some great items for our silent auction, and between the Trivia questions and the Minute-to-Win-It games, it's fun for all ages.

On December 3, 2015, I attended the PAH Advocacy Day at Queen's Park in Toronto. I was there, along with other advocates, to express our concern on a number of issues. Firstly, that Opsumit is still not available to patients who do not have private health insurance and who live outside of Quebec. Although Opsumit has been approved by Health Canada for use as a PAH medication, the Ontario government has yet to agree to pay for it. As a result, not all patients are able to access optimal treatment for their disease.

Secondly, we wanted MPPs to understand how patients' wellbeing could be negatively impacted by the proposed changes to how PAH drugs are prescribed. If CADTH recommendations are adopted by provinces, the Ontario government (along with other provincial governments) would enforce a "one-size-fits-all" approach to prescribing PAH medications—changes that would mean less doctor involvement in determining the most appropriate medication. I feel very strongly that it is important that it be my daughter's doctors that make the decisions regarding which medication(s) best suit her circumstances. If the Ontario government adopts CADTH recommendations, I am afraid that, in the future, when Brooke's health does deteriorate, her doctors may not be able to prescribe the best medication for her condition at that time. I fear that she will be given the "next one" on the list. That's what concerns me and that's why I went to Queen's Park.

Along with other advocates, I met two MPPs. We discussed our concerns about Opsumit and I told them Brooke's story and my concerns. They seemed genuinely interested, and seemed to understand that, while both of these issues might provide fiscal savings in the short term, they would eventually result in higher future costs in terms of more hospital trips and longer stays. At the very least, they heard the words "pulmonary hypertension," maybe for the first time, and learned a little about the disease.

The other thing our family did this year in regards to advocacy was to send emails to our Premier, Kathleen Wynne, our Minister of Health and Long Term Care, Dr. Eric Hoskins, and our MPP expressing our concern about these issues. It was so easy. We didn't even have to leave the comfort of our home. PHA Canada has a link to an already-written letter (**www.takeactionPAH.ca**). We simply had to add our name, enter our postal code and press send. It probably took less than a minute to send all three emails. We then asked extended family and friends to help us by sending the same emails to their MPPs. We were able to cover MPPs from ridings in Mississauga, Brampton, Woodbridge, Cornwall, Toronto, London, Oakville, and Ottawa.

I don't think one trip to Queen's Park will change anything. But I think it's important to keep trying and I have started on this journey. I realized on PAH Advocacy Day that politicians have to deal with many issues. A rare disease like pulmonary hypertension probably doesn't make their list of top ten things they need to worry about. If we want change to happen, we, both patients and caregivers, need to get the words "pulmonary hypertension" into the lexicon of our MPPs. We need to be a rare disease with a bigger voice.

And as luck would have it, we may have that opportunity here in Ontario. All the advocates who went to Queen's Park in December met an MPP from Kitchener-Conestoga, Michael Harris, who is making it his mission to bring more awareness of rare diseases to Queen's Park. Before the house rose for the holidays in December, he brought a motion forward to put together a Select Committee that will travel across Ontario to review treatments and funding for rare disease patients. He wants to meet and talk to caregivers and patients with rare diseases. He wants to understand the roadblocks—in diagnosis, in treatment, and in funding—faced by patients with rare diseases. What a platform for all rare diseases! What a platform for PH! I hope that by the time this text is published, the motion will have passed (it will be brought to the house for a vote on Thursday, March 3rd, the closest legislative day to Rare Disease Day, which is February 29, 2016). In the meantime, MPP Michael Harris is hoping to have as many people as possible who are impacted by rare diseases filling the Legislative chamber on the day the motion goes to a vote. I'll be there as I continue my journey as PH Advocate.

***If the Ontario government adopts CADTH recommendations, I am afraid that, in the future, when Brooke's health does deteriorate, her doctors may not be able to prescribe the best medication for her condition at that time. I fear that she will be given the "next one" on the list. That's what concerns me and that's why I went to Queen's Park.***

*Contributed by: Joan Paulin, PH caregiver and advocate, Mississauga, ON*


*Joan (left) helping out at the Trivia Fun Pub and Grub Night held in November 2015.*

# Tips and Tricks Shared from My Empowering SPIN Experience at the 16th Annual National Scleroderma Conference 2015

*Jeannie and members of the SPIN team.*

In September 2015, the Scleroderma Society of Canada (SSC) held its 16th Annual National Scleroderma Conference in Hamilton, Ontario. As a person living with pulmonary hypertension and pulmonary fibrosis secondary to scleroderma, I was eager to gain more knowledge of my complex health issues. This was my first time attending a scleroderma conference and networking with the community. Was I ever happy that I attended the event!

Present at Conference was the Scleroderma Patient-centered Intervention Network (SPIN), a research-centered organization supported by the Scleroderma Society of Ontario (SSO). SPIN is an organization that involves researchers, healthcare providers and people living with scleroderma from around the globe. SPIN's mission at Conference was to interview people living with scleroderma and physicians to record their testimonies through video. The organization aims to develop sets of non-pharmacological evidence-based resources that will improve quality of life for scleroderma patients.

SPIN invited me to contribute to their research by sharing a video testimony. They wanted to find out about the impact scleroderma had on my life, my goal-setting process, and how I establish a good rapport with my professional health team. I felt grateful for and empowered by this wonderful opportunity to share kernels of my journey, and by doing so assist others living with scleroderma.

Here are some transcribed segments of my video interview, through which I share my tips on how to set goals for oneself.

## GOAL SETTING TIPS AND TRICKS

Goal setting is important in our lives, although for people suffering from a chronic disease like PH or scleroderma, it can be a difficult thing to do. Approach the process of setting goals with determination and perseverance. To do so, think about a benefit you wish to achieve or a purpose you wish to attain both short and long term.

***To Achieve Your Goals, Think SMART...***

Be sure your goals are specific, measurable, and action-oriented. In planning out your goals, be resourceful and keep in mind time and frequency. Make sure that you can achieve them with, at a minimum, a 70% success rate.

Record your goals in writing. It is important to state your goals using positive language. Keep records of your goals in prominent places to remind yourself of them; if you think about your goals, you're more likely to achieve them. Have faith! Believe in yourself!

***Here are some examples to demonstrate how this SMART strategy works:***

***Exercise Goal:*** *My goal for pulmonary rehab is to gain greater stamina and energy, and to feel less breathlessness upon exertion.*

***How I plan to achieve this goal:*** *Two times a week, at each pulmonary rehab session, I walk for forty minutes on the treadmill at 2.0 mph with no incline. I use pursed lip breathing and liquid oxygen set at a continuous flow of 4L/min. Using a personal oximeter, I monitor my oxygen saturation and heart level during the workout. My goal is to maintain target levels with a minimum of 88% oxygen saturation and less than 120 beats a minute, with exertion.*

*The progress may seem slow, though each step brings us a step closer to the goal. Change takes time. Celebrate your success!*

***Positive Attitude Goal:*** *Maintain a positive attitude to deal with life's challenges.*

***How to achieve this goal:*** *Keep in mind that while we cannot always control what challenges are sent our way, we can control our reactions in dealing with them. Focus on the aspects of life that you can control. It is important not to let your medical diagnosis define who you are. You are more than your illness. Be grateful for the positive things in your life and the past challenges that you have overcome. Keep active in activities you are capable of doing and that give you joy!*

*Seek out caring family and friends in your support network. Focus on those who emit positive energies with a joy for living. Delegate responsibilities; people are willing to help, but need direction from you.*

*Keep a journal of your feelings and your journey. Attend and participate in support group meetings, where others understand much of what you are going through and can offer coping strategies. Be proactive by sharing your story with interested individuals and groups to promote awareness and advocacy.*

*Be thankful for each day... it is a gift! Live life to the fullest!*

***Ongoing Education Goal:*** *Having a chronic disease requires us to learn as much as we can to self-manage the symptoms to lead a manageable life.*

***How I plan to achieve this goal:*** *In addition to the information I get from my healthcare providers, I frequently seek out health talks at Patient and Family Education Libraries in hospitals, and attend conferences and workshops. I also gain a lot of information from listening to archived recordings from conferences and webinars that can be found on many reliable websites related to hospitals and organizations supporting various diseases.*

*con't next page*

Your Stories

*Another way to achieve this goal is to "pay it forward." After I have researched a topic of interest thoroughly, I share the information with others who are interested and with support groups. I also participate in Virtual Patient Groups at University Health Network (UHN) in Toronto several times a year to share my feedback on services. Recently, I have eagerly participated in the Very Important Patient (VIP) Program and Patient Partner Experience through Wightman-Berris Academy to help train UHN medical students to develop their clinical interview skills. I also stay involved with the community by sharing my story in the media, organizing events, and contributing articles.*

*When a stone is thrown into the water, its ripples carry on and on. With ongoing education about scleroderma and PH, we can motivate others by advocating and sharing our stories with our challenges and coping strategies.*

*Contributed by: Jeannie Tom, PH and scleroderma patient, Willowdale, ON*

**RELATED WEB LINKS:**

Scleroderma Society of Canada: www.scleroderma.ca

SPIN: www.spinsclero.com

Medical Minute

# Ask a Nurse: Emergency Kit

Expect the unexpected. Words to live by particularly if you have any health-related issues. All individuals with pulmonary hypertension should be prepared for an emergency or unexpected event. Keeping an emergency kit at hand is a helpful way to ensure you have all of your health information as well as everything you might need when one of these unexpected events arises. Keeping everything together in a box, case, or tote makes it easy to locate your emergency kit material when you need it.

## EMERGENCY KIT ESSENTIALS

You should always carry a list that includes the phone numbers of: all your physicians, specialty pharmacies, oxygen provider, local emergency phone numbers, as well as the name and number of your emergency contact (the person to notify in the event of an emergency). When travelling, it is a good idea to find locations and phone numbers of health care providers specializing in pulmonary hypertension where you are travelling.

You should also include an up-to-date list of all your medications including the name, dose, and how often you are taking the medication. This list should include all prescription medications, medications you purchase from your drugstore or pharmacist, vitamins and supplements, as well as the flow rate of your oxygen. In addition, it should include a list of any medication allergies you may have. Your PH specialist may change your medications or the amount of medication that you take, so ensure that you update this list every time a change is made.

It is important to have a copy of these two lists with you at all times. Your health care providers can assist you with keeping the lists accurate and up to date.

## MEDICATION AND SUPPLIES

Be sure to take your medications, in their original bottle, with you whenever you travel or go to the hospital. Not all pharmacies, including hospital pharmacies, may carry your medications. Keeping all of your medications together in one place, whenever possible, makes it easier for you to take them when you need to, and easier for someone else to locate them if necessary. If you are receiving your medications through a continuous infusion, your emergency kit should include any medical supplies required for inserting, mixing, and infusing your medications; line and infusion site care; and a portable cooler with ice packs. For individuals who are on oxygen, be sure to have enough tubing, tanks, and regulators on hand at all times. These supplies need to go with you as well when travelling or going to the hospital.

## USEFUL ITEMS

It is a good idea to wear a Medic Alert bracelet or necklace. Your medical team or local pharmacist can help you order one. You may request a brief note from your physician to carry with you as well, which explains your illness and medications. This is particularly important if you are receiving any of your medications through a continuous infusion, which many pulmonary hypertension clinics have already prepared.

Other useful items to have in your emergency kit are a blood pressure monitor and thermometer. These can provide important information when speaking with your health care providers. Many of the medications prescribed to treat pulmonary hypertension can have an effect on your blood pressure and a fever is one of the signs of infection.

Being prepared can help avoid added stress when dealing with an unexpected event or emergency.

*Contributed by: Brenda Bunting, Nurse Coordinator, Southwestern Ontario Pulmonary Hypertension Clinic, London, ON*

***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.***

# The Role of the Nitric Oxide Pathway in PAH

In this issue, we will address some questions regarding PH therapy, more specifically, the role of the nitric oxide pathway in PAH and the drugs targeting this pathway.

### *What is the nitric oxide (NO) pathway composed of?*

This pathway is one of the crucial pathways leading to blood vessel relaxation (vasodilation) in the lungs. Nitric oxide (NO) is produced from a molecule called L-arginine, an amino acid that can be synthesised in our body or acquired from diet, with the help of nitric oxide synthase (NOS). NO binds soluble guanylate cyclase (sGC), leading to production of a new molecule called cyclic guanisone monophosphate (cGMP). cGMP is the molecule responsible for vasodilation/relaxation of the blood vessels; however, it can be degraded by a family of enzymes called the phosphodiesterase-5 (PDE-5). NO is usually produced in endothelial cells—the cells lining the inside of blood vessels—but then is taken up by smooth muscle cells—the cells making the second layer within a blood vessel that is important for vessel contraction.

### *I am confused! How does this pathway work again?*

To summarize:

- High levels of NO/sGC result in high production of cGMP, leading to blood vessel relaxation.
- Low levels of NO/sGC result in low production of cGMP, leading to blood vessel tightening.

### *Why is this pathway important in PAH?*

In PAH, there are dramatic decreases in NO. This results in low vessel relaxation, and constant constriction, leading to narrowing of the blood vessels in the lungs. The effect of this is increased pressure in the lungs and pulmonary arteries.

### *What treatments target this pathway in PAH and how do they work?*

Therapies attempt to enhance this pathway in order to increase vessel relaxation. The first class of drugs inhibit PDE5, which means cGMP is sustained and not degraded, resulting in blood vessel relaxation. The other class of drugs act to stimulate sGC function and its binding to NO, leading to increased cGMP production.

### *Currently, which therapies targeting this pathway are approved for PAH?*

Sildenafil (Viagra, Revatio) and Tadalafil (AdCirca, Cialis) are PDE5 inhibitors approved for treatment in PAH patients in Canada since 2006 and 2010 respectively. Recently, a sGC stimulator, Riociguat, was approved for PAH (WHO class I, 2014) as well as patients suffering from non-operative or residual post-operative chronic thromboembolic pulmonary hypertension (CTEPH WHO class IV, 2013).

### *What does research show about these drugs? Which one is better?*

Some side effects can appear with using these drugs. For the PDE5 inhibitors, there have been reports of headaches, flushing, muscle pain, and indigestion on the short term, which can develop into diarrhea and peripheral edema in the long term. Stimulation of sGC has also been linked to similar side effects on both short and long term, accompanied with possible hypotension and vomiting. One cannot say that one drug is definitely better than another since each patient is affected differently by the disease and will have a different response to drugs. Thus, it is very important to discuss with a PH specialist which drug is most appropriate for each patient.

**REFERENCES:**

*1) Provencher and Granton. Current Treatment Approaches to Pulmonary Arterial Hypertension (2015). Canadian Journal of Cardiology.*

*2) Humbert and Ghofrani. The Molecular Targets of Approved Treatments for Pulmonary Arterial Hypertension (2015). Thorax.*

*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.*

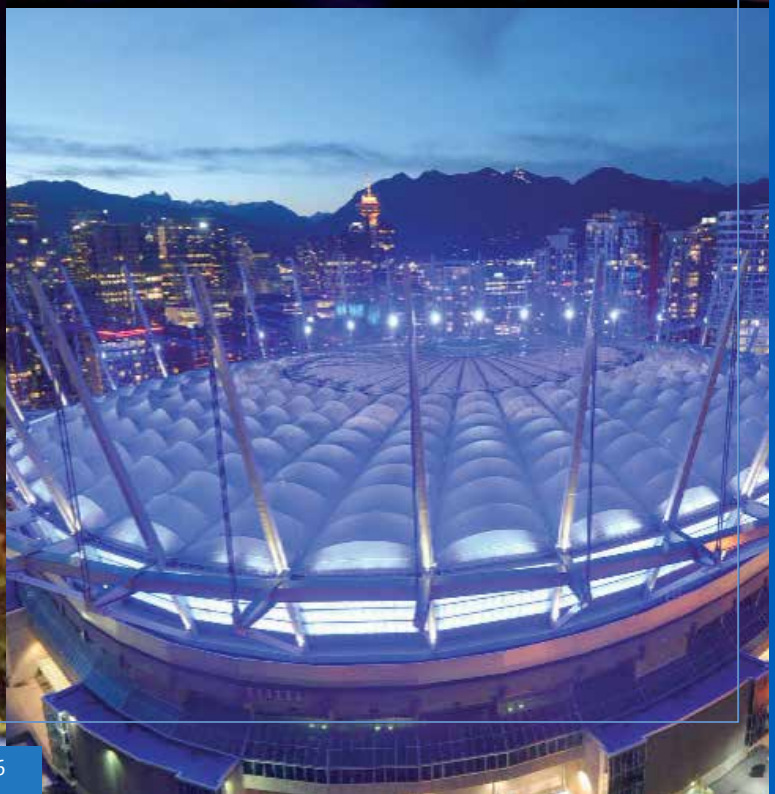
# Help us Paint Canada Purple for World PH Day!

Thursday May 5th, 2016 marks the 5th annual World PH Day. Again this year, PHA Canada will celebrate and spread awareness of PH by illuminating several Canadian Monuments in Purple. In 2015, thanks to the efforts of members of our community, 14 monuments were illuminated in periwinkle from shore to shore including landmarks such as: BC Place, Vancouver; Calgary Tower, Calgary; High Level Bridge, Edmonton; CN Tower, Toronto; Ottawa Heritage Building, Ottawa; and Olympic Tower, Montreal!

To ensure that Canada shines bright in periwinkle on May 5th, please help us by sending requests for monuments in your area to be illuminated. PH patient and advocate Loretta Chu has researched an impressive list of Canadian monuments that accept illumination requests. To access this list and to get tips on how to present a request, please visit **www.phacanada.ca/PaintCanadaPurple**.

We look forward to seeing Canada shine bright on World PH Day!

**#PaintCanadaPurple**



# In Memory 2015

While our community holds enormous hope for the future, the reality is that pulmonary hypertension still takes loved ones away from us. The success stories from our members, developments in research, and heightened awareness about PH are worth celebrating, but they cannot bring back the people that we have lost to this disease. The following individuals are those who sadly lost their battle to PH over the 2015 year. Our hearts go out to these individuals and their families.

***Brenden Brinkworth***
***Justin Bourassa***
***Carly St-Aubin-Chartrand***
***Wilna Burboyne-Toombs***
***Denise Filiatrault***
***Christine Gougeon***
***Joanne Grenier***
***Brenda Jonk-Andries***
***Lila Klussmann***
***Jeannine Lafrenière***
***Ghislaine Mongeau***
***Tony Passarella***
***Renald Perée***
***Margaret Petrie***
***Renée Prieur***
***Ada-Menucha Riven***
***Blaž Varjaci***
***Dawn West***
***Jane Williams***
***Letisha Winning***
***Jacky Yang***
***Robert Yasky***
***Kristy-Lynn Zizian***



# 2015 Donor Recognition

## CORPORATE EFFORTS

The PHA Canada Corporate Committee members support the mission and vision of PHA Canada via a yearly dues contribution. Corporate Committee members in 2015 were:

***Actelion Pharmaceuticals Canada***
***Bayer Inc.***
***GlaxoSmithKline Inc.***
***McKesson Specialty Health***
***Pfizer Canada***
***Shoppers Drug Mart Specialty Health***
***Unither Biotech Inc.***

## DONORS

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 all of those who contributed in any way in 2015. We appreciate each dollar donated, as 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.

### $500 – $999

***Amin Adatiya***
***Pepy Cordick***
***Elaine Goodhue***
***Pat James***
***Peter Littlejohn***
***Fuller Utility Services***
***North Waterloo Farmers Mutual Insurance***
***Duncan Stewart***
***St-Germain and Brinkworth families***

### $1000 – $4999

***Finch and District Lions Club***
***Sandra Langford***
***Sanjay Mehta***
***John Murray***
***Scotia Bank (Russell, ON)***
***Sharon Proudfoot***
***Edward Rathonyi-Reusz***
***Quentin Smith***

***Glenn Watt (The Tenaquip Foundation)***

### $5000 +

***Bell Family***
***Blanchette Family***
***Phil Paroian (Paroian Family PHA Canada PH Research Scholarship)***

## 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 for PH*** *(London, ON)*
***Brooklin Quiz Night*** *(Brooklin, ON)*
***Cornwall Stride For Breath*** *(Cornwall, ON)*
***GolPH for PH*** *(Brampton, ON)*
***Lend a Hand for PH 2015 campaign***
***Marche à la mémoire de Carly*** *(Gatineau, QC)*
***Marion Feeley fundraiser*** *(Russell, ON)*
***Pierce Family fundraiser*** *(West Lorne, ON)*
***Second Annual PHA Canada Run/Walk for Research*** *(Ajax, ON)*
***Second Annual Run/Walk, Swim or Bike/Spin PH Challenge*** *(Bradford, ON)*
***Third Annual DINE for the CAUSE and Silent Auction*** *(Ottawa, ON)*
***Third Annual Masquerade Ball*** *(Calgary, AB)*
***Trivia Fun Pub and Grub Night*** *(Oakville, ON)*

# *Connections* submission guidelines

The deadline for submissions for the next issue of *Connections* is July 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:**

Subject: *Connections* submission

**connections@phacanada.ca**

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.