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

Message from the Chair of the Board

Hello again PHriends,

2015 is already shaping up as an exciting year for the Canadian PH community. Our 4th National PH Conference in Montreal May 1-3rd is rapidly approaching! Whether you are a veteran conference attendee or if this will be your first, going to Conference is the best way to feel part of our amazing PH community: you will find kindred spirits in other PH patients and caregivers who are going through the same thing you are. You will also learn about the newest PH research and treatments, and how to get involved in advocacy efforts. See the article in this issue of Connections by Bronwyn McBride on why attending Conference is so important.

At PHA Canada, we continue to work hard in our commitment to support PH patients and their caregivers, and remain focused on our vision of a better life for all Canadians affected by PH. PHA Canada has just defined our first ever 5-year strategic plan. This plan recognizes 5 key issues facing PH patients and caregivers today in Canada, and addresses each with specific goals over the next 2-5 years. Advocacy on behalf of the Canadian PH community is one of PHA Canada's 5 key priorities, as PH is a rare disease, often ignored by government, and for which medications are exceedingly expensive.

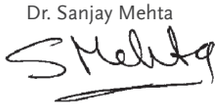
Advocacy can take many forms: see the article by Dr. David Ostrow of the Vancouver PH Clinic. Individual patients often have to learn to advocate for themselves while negotiating our healthcare system, as for example in dealing with their own family doctor, as Lisa Lee explains. PH patients can also advocate for others, as described in the article by PH patient and PHA Canada Ambassador Tarya Laviolette. Individuals like you can have a big impact on government and other organizations, which are often most responsive to the voices of those directly affected by an illness. Even just sharing your story with media and the public helps improve general awareness of PH in Canada.

When faced with a diagnosis of a serious illness like PH, it is easy to want to crawl into a corner and hide from family, friends, and especially strangers. However, sharing our stories is part of what makes us all human. Connecting with others lessens the burden of dealing with illness, as we realize that we are not alone! Traditionally, women are more willing to share stories and feelings as they go through the PH journey. However, more men are standing up to tell their PH stories than ever before, as seen in this issue of Connections! Regardless of their gender, each patient's story is worth sharing, and we're excited to have such wonderful contributions.

PHA Canada is proud to have already improved the lives of Canadians affected by PH, but we think we can offer our patients and caregivers so much more. To do this, we need your help. See Vice-Chair Roberta Massender's article on one of the most important things you can do to support the PH community. I look forward to seeing many of you again at our Conference in Montreal, and meeting some of you for the first time, as we all work together as part of the growing Canadian PH community.

With hope,

Dr. Sanjay Mehta



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In This Issue:

My daughter, my rock – the story of our joint family

PH patient Mechelle (on the right) with her caring daughter Mandy.

"Mandy is my daughter (I don't think of her any other way), and my best friend. We laugh together and cry together, and I couldn't have asked for a more loving and wonderful caregiver."

Where to begin? My name is Mechelle Ramdeen. In February 2007, I was newly married to my husband Dass and had my son Taidyn. I was diagnosed with idiopathic pulmonary hypertension when Taidyn was only fourteen months old. In 2008, our family faced another change: my stepdaughters Mandy, 17 years old, and Tina, 13, would soon be moving in with us.

Mandy and Tina had lost their biological mother to health related issues when they were young children. Because Mandy and Tina had experienced this tragedy, I was devastated to feel that they were now becoming a part of my life, and I was very sick too. Everyone knows the story of Cinderella and the wicked stepmother - I vowed that I was not going to be like that, and that we were going to be a family. Well, let's just say that I got a lot more then I bargained for.

Mandy is my oldest stepdaughter who is now 23. Having her is so much more then I could have ever imagined when I first became a stepmother. She is my best friend and rock on the good days, and is my caregiver on the bad ones. Although I try not to have many, there are days where I can't get out of bed or I just need that little extra rest to be good for the day. Mandy looks after the family for me and gets meals ready. She even sees to her little brother Taidyn, now 8 years old, by getting him off to school or sports events. Mandy holds our household together when I am too sick to do so.

Even more importantly, she always looks after me. Whether it's the simplest things, like getting my pills, or the tough stuff, like helping me just to function, Mandy is there by my side. She drives me to all my appointments with my doctors when she is home. She has seen the best and the worst possible of this awful disease. Mandy has also befriended our PH family, and has grieved with me when I lose friends to PH. She is never far from me: wherever I am, she and Taidyn are close behind.

Mandy is currently attending a Personal Support Worker program which is going to be bridging into a Registered Nurse Practitioner program. She also works as a customer service manager in Walmart. However, her work often doesn't end there: on many days when she comes home, there's a lot of work to be done around the house, too. Because of my illness, Mandy adopts my role by doing everything that a mother would do for her family.

Mandy is the type of girl that wears her heart on her sleeve: she would do anything to help out a friend, no matter what. She just smiles, no questions asked, and does whatever it takes. Our relationship goes far beyond the role of stepmother and stepdaughter. Mandy is my daughter (I don't think of her any other way), and my best friend. We laugh together and cry together, and I couldn't have asked for a more loving and wonderful caregiver. She is my rock, and one day she's going to make the best nurse possible: I know this for a fact, because she is the best nurse for me. I love you so much Mandy, and hope that you follow your heart and dreams!

Contributed by: Mechelle Ramdeen, PH patient, Vittoria, ON

From swimming to sinking: my story

I am a patient who was diagnosed about 10 years ago with pulmonary hypertension.

In 1997, I was swimming across a small lake in northern Ontario with one of my daughters. I had been doing this for a number of years successfully with my children. We took a small break on the far side of the lake, turned around, and swam back. This particular time on my way back, I was about two-thirds of the way and suddenly I couldn't breathe. The lake was as calm as glass, so I wasn't fighting the current in any way: it was a very relaxed swim. I could not call for help! It seemed like an eternity before one of the men on shore saw that I was in trouble. My young neighbour jumped in and pulled me to safety. By now part of my shortness of breath was from fear and the other part, I had no idea, but I didn't think I had worked myself up to that degree of breathlessness with fear alone. An ambulance was called and I was immediately put on oxygen and hospitalized for three days. When discharged and I got back to the lake, I made a point of going for a swim. I never wanted the fear to stick with me as I have always enjoyed swimming.

Shortly after that episode, I was referred to a cardiologist and tests and medications were started. I was diagnosed with congestive heart failure.

I was travelling from Burlington to Toronto to work every day, and for some reason the stairs became more and more difficult for me to manage. I was so out of breath and energy, having to take a break after every 3 – 4 stairs to catch my breath. The walk up underground from the train to my desk in an office tower was taking me longer and longer: I was exhausted before my work day started. The inclines and declines in the underground would mean nothing to a healthy person, but to me, they were very difficult, as was the opening of the big heavy doors between buildings. I couldn't understand what was making me so out of breath. I hoped every time that I wouldn't meet anyone who wanted to talk, as talking and walking at the same time were out of the question. The overwhelming exhaustion in me was something I'd never experienced, as I'd always had high energy. I just thought I was going through the winter doldrums until I found myself sitting straight up on the side of my bed, struggling to get enough air. I was taken to the hospital again, and this resulted in a referral to a cardiologist. Tests were done and a regime of medication was changed, but I continued to be very short of breath.

I was very full of fluid most of the time, and swelling all over even after the episode of congestive heart failure. This too was contributing to my shortness of breath and exhaustion. I was referred to a nephrologist who diagnosed me with chronic kidney disease, and another regime of medications were prescribed to reduce the fluid retention.

After a review of my file, my team of doctors at Oakville Trafalgar Memorial Hospital decided there was something else getting in the way of a clear picture, and I was referred to a lung specialist at the hospital. He started me on an inhaler and soon after, he referred me to a specialist in pulmonary hypertension. I had never heard of pulmonary hypertension and found the unfamiliar name intimidating. I looked it up on the internet and found I was even more intimidated, and decided to wait for information from the specialist I had been referred to.

cont'd on next page

Barb (rightmost) and friends, who are other PH patients and caregivers, at January's Ontario Lung Association 'Breathe' Gala.

My referral was to a pulmonary hypertension specialist in London, Ontario where I was finally diagnosed with pulmonary venous disease as a result of left-sided heart disease, which is one of the more common types of pulmonary hypertension. I have mitral valve regurgitation and atrial fibrillation, and also have asthma. A lot of patients are misdiagnosed with asthma when in actual fact they have pulmonary hypertension, but I have both. After these diagnoses, I was put on Long Term Disability.

PHA Canada's Toronto chapter have partnered with the Ontario Lung Association, along with numerous lung stakeholders through a Lung Health Alliance. Together, they are developing a Lung Health Action Plan which they hope will be adopted by the Province of Ontario to improve the lives of all patients with lung disease. This type of advocacy gives me a purpose and hope that in the future, others with lung conditions will receive better care.

I give sincere thanks to a wonderful medical team, supportive caregivers, many friends and family including the members of the PHA Canada Toronto Chapter Support Group. They are always available for support to patients, caregivers, friends and family, through telephone, e-mail and our regular meetings. You are all truly amazing, and my phriends are the one thing I am thankful to PH for.

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

How I learned to cope with the emotional toll of my PH diagnosis – taking a dose of my own medicine

As I am sure many pulmonary hypertension patients can identify with, when I was diagnosed, life as I knew it took a nosedive. My accompanying emotions were all over the place: I was relieved that I finally knew what I had been suffering from since childhood. I was sad and angry that I had to leave my home and abandon my goals. I was afraid that I may die before I had finished doing everything I wanted to do. Finally, I was frustrated that I could not 'fix' myself. Eventually, I found myself in a situational depression. The feeling of hopelessness and loss of independence was crippling. There were some hopeful moments and decisions that helped me to move forward, but overall, my emotional theme was gloomy.

Today, this picture is very different. I am very positive and hopeful overall, which I attribute to many different things - incredible medical care, improvement in my condition, re-gained independence and of course the selfless perseverance of wonderfully supportive people in my life, to name a few. However, I would like to write today about how my own knowledge of a counseling skill set called Cognitive Behavioural Therapy (or CBT) has helped me to work through many of my strong and troubling emotions. I share this because, though this process takes commitment and personal work, anyone can do it!

I should also tell you that I am a Social Worker. Most recently, I've worked as a clinical counselor in a Nurse Practitioner-led clinic helping people

> ***"The feeling of hopelessness and loss of independence was crippling."***

through everything from mental health to chronic illness diagnosis to addictions and poverty. CBT is a skill set I use in this work.

CBT essentially refers to how our thoughts, feelings and behaviours are connected and how by manipulating one, the others are affected. CBT counseling can help folks learn to control and change how they think and or behave, which affects their emotional state in a positive way.

'The underlying concept behind CBT is that our thoughts and feelings play a fundamental role in our behavior. For example, a person who spends a lot of time thinking about plane crashes, runway accidents, and other air disasters may find themselves avoiding air travel. The goal of cognitive behavior therapy is to teach patients that while they cannot control every aspect of the world around them, they can take control of how they interpret and deal with things in their environment' (Cherry, K. in psychology.about.com)

To reference a popular children's book called 'The Little Engine That Could', the slogan "I think I can" sums up the concept of CBT. However, the practice is a bit more complex and takes more work to accomplish. If attainable, seeking professional guidance is recommended to ensure success. If this is not easily attained, a good alternative is learning through some of the relevant literature. I suggest Mind Over Mood: Change How You Feel By Changing How You Think, a CBT workbook by Dennis Greenberger

Emily acting as a speaker at PHA Canada's 2013 National PH Conference in Ottawa, ON.

"I realized I had to live with myself and quite frankly; I did not want to live with a negative person. Something had to change."

and Christine A. Padesky (1995) for a user-friendly self-help version of Cognitive Behavioural Therapy. The first step is becoming more aware of your thoughts and particularly 'hot thoughts': judgmental thoughts that are generally absolutist, irrational and emotionally driven.

This is an example from my own experience. I used to look at others doing things I could not (let's say, running) and think things like "I'll never be able to do that", "it's not fair", "it must be nice to be able to run". My corresponding mood would be sad, angry, jealous, and feeling sorry for myself. The problem with my thoughts was that they were polarized, meaning that I had an all or nothing mentality. They were also irrational: I don't know for sure that I'll never be able to run. My thoughts were also discounting of others' struggles and my strengths. I didn't know what that person might be struggling with, and I was only focusing on what I could not do and not what I could do. I have strengths and weaknesses, just like any other person. And finally, my thoughts are negatively framed. I was mentally filtering out the positive and choosing to focus on the negative. We humans tend to do this. It is called a cognitive distortion; a fancy term for how our minds like to rationalize irrational thoughts and convince us that something that is untrue is true. For example, thinking "I am useless" can be a belief that we then reinforce with behavioural patterns that reinforce the belief, even in the face of contradicting evidence.

The good news is that by becoming aware of our thoughts (distorted and otherwise) and how they correspond with our behaviours and feelings, we can condition our brains to think differently. This can help us to feel and behave differently.

I personally began this journey when I had had enough of my own gloominess. I realized I had to live with myself and quite frankly; I did not want to live with a negative person. Something had to change. I began to challenge my own thoughts and purposefully changed my behaviour: I went back to school part-time and via distance and finished the degree I had been working on before I became ill. For me, this was significant because I was acknowledging a strength – my intellectual capacity and social work skills. This I could do, so why not do it! With this action, I began to feel better: not every day, but most days; more than before. It's important to measure even incremental change. Try doing this by keeping a record of your moods and rate them, for example "Feeling sad this morning, 8/10 (10 being the worst and 0 not sad at all), lasted 2 hours". Note the corresponding behaviours and thoughts, even small changes like smiling at people, or thinking about how lucky one is for the support people in their lives. Thinking about these positive things helped me to become more cognizant of how my illness actually brought me closer to my loved ones. I chose to behave in a way which expressed this gratitude. This can help in motivating us to continue to engage in behaviour that helps us feel better.

A good way to start this CBT process is to start a diary. Write down your thoughts, feelings and behaviours and note how they correspond. Try and spot the cognitive distortions and write down alternatives when you notice patterns of negativity; ways that you can look at the situation differently. Do this consistently and these new ways of thinking, behaving and feeling will eventually become second nature and habitual.

It will not always be easy, and you will always have the odd rough moment. You are human, after all, and perfection is not attainable! However, building these skills is worth it. You will increase your resiliency and truly be able to enjoy what you have. I kind of love this positive person I now live with; she is pretty awesome!

Contributed by: Emily Dolan, MSW, RSW, PH patient, Toronto, ON

Emily (leftmost) surrounded by friends and family, all of whom are great supporters of PHA Canada.

Dealing with depression in chronic illness

The Blues, melancholy, the Black Dog: depression is known by many names. Its association with chronic illness is well known, yet the symptoms of depression may be overlooked as being part of the illness. It is not caused by personal weakness or having "a bad attitude". Being depressed does not mean that you are "crazy". When your body betrays you, it is natural to feel sad, angry or hopeless. The reality is that pulmonary hypertension patients and their family members are all affected by the disease in emotional ways. We must mentally let go of the future we thought we would have, grieve for its loss and face a new reality; one with daily challenges.

Some symptoms of depression can be a loss of interest in usual activities, hopelessness, fatigue, change in appetite, sleep disturbance, increased aches and pains, sadness, irritability or anger.

Talk to your GP about your symptoms. They may refer you to a psychologist or therapist. It's important to remember that not all counsellors are the same, and you may have to try a few before you find one that you feel comfortable with. My GP was most helpful. He explained that sometimes you can be depressed for so long that you forget what normal feels like. If depression is severe or long term, chemical changes take place in the brain. Anti-depressants may be necessary to restore the balance.

"PH can be an isolating illness as our worlds can become much smaller. Remember that you are NOT ALONE."

There are many strategies available to help with depression: you can join a PH support group, try gentle exercise, do yoga, meditate, listen to music, create a restful place in your home to relax, practice gratitude, seek things to look forward to, learn mindfulness, try breathing techniques and learn stress reduction exercises. Eating healthier and finding ways to make daily activities easier will lessen stress. Time management is a very useful tool, as it helps you to avoid overdoing activities one day at the cost of your energy for the next. Exercise regularly to release your body's natural endorphins.

PH can be an isolating illness as our worlds can become much smaller. Remember that you are not alone. The internet is a great way to stay in touch with friends and reach out to other people living with PH. Find the number of your local Crisis Line if you feel like talking to someone immediately. In a mental health emergency, your local emergency department will get you the help you need.

In my case, medication, along with counselling and exercise have made a huge difference. For two years, I could not use my art studio. My creativity disappeared when the depression took hold. Now I am back in my studio and writing again.

The Black Dog accompanies me still
But my arsenal contains more tricks
To make him sit and stay.
I chip away at my protective armour
Until it yields.
Each piece that falls
Makes the world a brighter place.
I emerge, a new resilient self
And step forward, towards tomorrow.

Contributed by: Jill Morton, PH patient, Victoria, BC.

Adventures in advocacy

Research versus advocacy. I've heard a lot about these two concepts recently, and I think it makes for interesting discussion. Where should we, as the PH community, be focusing our efforts? How should we be spending our money? Why spend our resources talking, creating petitions and meeting with our local government bodies when what we really need is a cure? What does the government have to do with my medical treatment, anyway?

A lot, actually.

When I was first diagnosed, my doctor told me I had two treatment options: Flolan (IV therapy) or a combination of at least two oral drugs. As a newly diagnosed patient, I was pretty frightened of the IV drugs at the time, so I jumped at the chance to try oral medication. There was just one catch – combination therapy at the time of diagnosis isn't funded in my province. The way PharmaCare works in BC, I would have to start on one drug, get worse, and then apply for funding to add a second drug. I asked if I could simply pay for whatever wasn't covered by the province... and then I heard the price tag.

I was lucky enough to have a third option. My PH clinic was involved in a clinical trial that was studying the effects of combination therapy at diagnosis. I decided to join the study instead of going on IV meds, even though I only had a 50% chance of getting both drugs right up front. I got lucky, and was given both drugs at the same time; I was also fortunate enough that I responded really well to the medications, and my test results look very good. 'Stable' is my new favourite word.

Incredible Advocate and PHA Canada Ambassador Tarya.

The fact remains, however, that newly diagnosed patients in my province don't have access to the treatment I got by joining up with that clinical trial. That study is now over, and the results showed huge benefits for people like me, diagnosed in class three, starting off with a combination of oral drugs.

This is why I do advocacy work; this is why I talk to my government representatives. What I want is pretty simple. I want PH specialists – the most experienced, informed and well-trained physicians in the field of pulmonary hypertension in Canada – to be the ones who decide on the treatment plan that is right for Canadians faced with this rare disease. While these specialists are able to prescribe the treatments they think are appropriate, they don't have the power to make decisions about funding. Drug funding decisions are made at the provincial level of government, so it's those representatives we need to be talking to. Chances are, they've never heard of pulmonary hypertension, and they have no idea of the impact of this disease and of timely access to appropriate treatment on our lives. Until we tell them.

"I want PH specialists – the most experienced, informed and well-trained physicians in the field of pulmonary hypertension in Canada – to be the ones who decide on the treatment plan that is right for Canadians faced with this rare disease."

While doing research into new PH treatments and eventually a cure for PH is critical, the reality is that right now, many Canadians can't even access some of the best treatments available. **This is why I'm glad that PHA Canada is fighting to make sure that all Canadians living with pulmonary hypertension have access to whatever treatment their PH specialist thinks is appropriate.** The research happening in PH right now is amazing, and I want to use my story, my experience, my voice to ensure that our government representatives, the gatekeepers to our access to new medications, truly understand what these new treatments mean to us. I invite you to join me in this fight. Let's make our voices heard.

Contributed by: Tarya Laviolette, PH patient, Vancouver, BC.

Effective advocacy — a physician's perspective

Advocacy for rare diseases—those that do not attract the public eye, or that of policy makers—is crucial. Working through the media can bring a rare disease like pulmonary hypertension to light. Based on my experience, I have outlined some of what I hope are helpful clues to make that advocacy successful.

- You need a "hook" to snare the attention of both media outlets and policy makers. That hook is a story about a patient (or caregiver). I favor a positive story, and we know that with many new treatments for PH, there are positive stories. Embedded in the story, there needs to be "the ask"—the thing that you want the individual to do.

- You need to make sure that the policy makers see themselves as partners in a successful partnership. Wrap "the ask" around what the patients and their families have already done to improve the life of PH patients. Don't be afraid to quantify the work done, and also outline what other professionals, (physicians, nurses, etc) have done, including time they have spent outside their traditional roles. Then, "the ask" doesn't seem so big in comparison, and it will be tagged to a successful partnership. Before we ask politicians and others for money for research, what have we done ourselves to support research and/or care? For patient organizations doing advocacy, it might be to partially fund a local or national investigator, or an educator-training program, for example.

Dr. David Ostrow.

- Be specific about "the ask": be able to say with confidence how it will contribute to the good news story that you have already created.

- Demonstrate your links to other groups. In the case of PH, the logical association is with the international community of PH associations and with the other national associations of rare conditions. We did a joint presentation to parliamentarians with CORD (the Canadian Organization of Rare Diseases) and the assistance of some pharmaceutical companies a number of years ago. This activity and partnership needs to be regular and relentless.

- The pharmaceutical industry is your partner. Sure, they benefit when drugs are approved for treatment. However, they do the research, and along with the academic centers, they support the clinical trials that get patients on new treatments early. They also contribute, often generously, to different patient support programs and education programs.

- Never say "there is no money for our condition". There is always money being spent: we just need to leverage that spend and show how "just a bit more" will get a successful outcome.

Finally, to slightly modify an age old quote: "If we are not for ourselves, who is for us? And being for ourselves, what are 'we'? And if not now, then when?"

Contributed by: Dr. David Ostrow, MD, FRCPC, Vancouver, BC.

Roberta (centre) has been involved with the PH community for 15 years, ever since her niece was diagnosed. Here she smiles with Jennifer Gendron (left) and Angie Knott (right) at the PHA Canada 2013 National PH Conference.

On the importance of membership

Where my PH story began

My life was touched by PH when my niece, Nicole Harrison, was diagnosed with pulmonary hypertension. In November 2000, Nicole was admitted to Vancouver General Hospital, her Hickman catheter was inserted and Flolan titration began. Nicole lived every day to the fullest and was an inspiration to me and all of those around her, a true champion. I learned so much from Nicole as our family took the PH journey with her. Sadly, Nicole lost her fight with PH and died on March 18, 2006.

Many things have changed since Nicole was diagnosed. Then, there was only Flolan or lung transplant, today there are 9 approved drugs with numerous combination therapies. Today, PH patients are not alone and they have more options and a brighter future than ever before.

From that day in November 2000, I have been a member and supporter of the PH community. I continue to be a part of this community in honour of Nicole's fight. We have more work to do: we need better treatments, better support for our patients, and we need to close the gap from onset of symptoms and diagnosis. And most important, we need to find a cure.

Many of us might not realize it, but becoming a member of PHA Canada is one of the greatest things a person can do to support the PH community.

What membership does for PHA Canada

PHA Canada was founded to create a better life for people with PH and the people that care about them. By becoming a member and supporter of PHA Canada, you stand as part of a community of hope that allows an opportunity to connect, learn and fight back against this terrible illness.

Without the support of our members, we would be unable to help in bringing hope and support into the lives of many patients and their families. A strong membership base helps ensure that PHA Canada can offer resources to those affected by PH – patients, family members, caregivers, friends.

When you become a member of PHA Canada, you are making a decision to give something back. Numbers give us strength and make our voice louder as we work toward a better life for everyone affected by PH. Perhaps individuals don't become members because they feel that they don't benefit, or it's not important to them. I would really like to emphasize that for PHA Canada, having more members benefits US, by making us a stronger organization. It helps our advocacy efforts, because the bigger a group of people we are, the more powerful we are. It also helps when facing the media. Did you know that one of the first questions the media will ask in an interview with PHA Canada community members is "How many people does your organization represent?" When we say "We have 450 members across Canada", we're really saying "this is how many people care about this disease." And that message can have a big impact.

How can I help?

You can make a tangible contribution to the PH community by becoming a member today. Membership is not only for patients and caregivers, but for family and extended family, the friend you meet for coffee, your co-worker, your neighbour, and anyone else who knows your story and wants to help.

Ask your family and friends to become members and supporters of PHA Canada. It is easy to do and becoming a member can be their gift to you.

All of us together, the staff and board of PHA Canada, patients and caregivers, families and friends, medical professionals and the corporate community will be a strong and powerful voice as we continue the fight against pulmonary hypertension. Thank you for your membership in our association.

Contributed by: Roberta Massender, Vice Chair, PHA Canada, President, BC Pulmonary Hypertension Society, Richmond, BC.

The male perspective on PH

It's well known in research surrounding pulmonary hypertension that PH affects a greater number of women than it does men. As such, many of the articles we feature in Connections magazine and in our other publications focus on women's stories of living and loving life despite the challenges of PH. However, there are many men who also live with this disease and go through a wide variety of experiences, from diagnosis to travelling and from family to community. The next few articles feature the personal stories of Canadian men who are PAH and chronic thromboembolic pulmonary hypertension (CTEPH) patients. These are their journeys, which illustrate that this disease can affect men—husbands, fathers, professionals and more—in unique ways. Thanks to each of these contributors for sharing their lives with us.

Your Stories

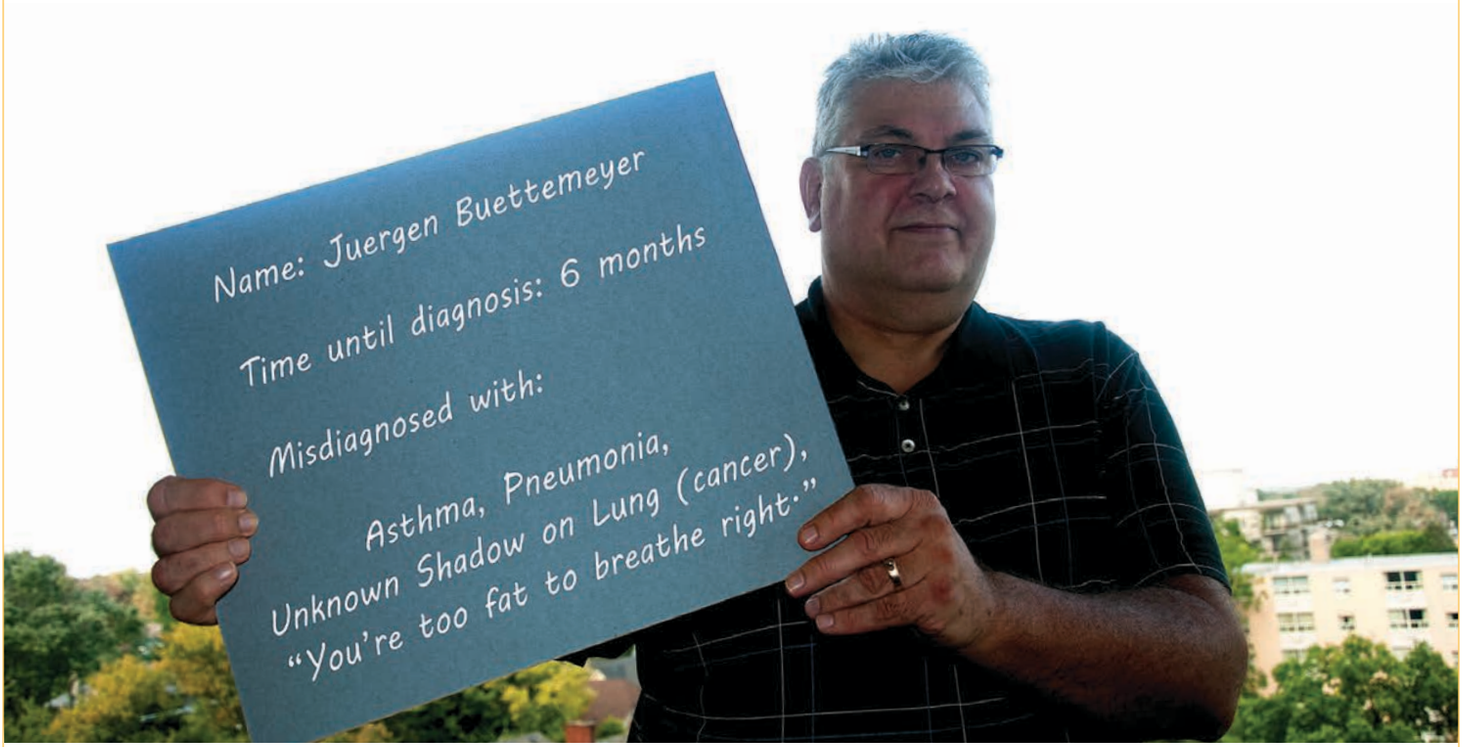
CTEPH – not quite like winning the lottery

After knee replacement surgery in July 2012, I developed shortness of breath when climbing stairs. The diagnoses I got when visiting my GP and the local ER were asthma, possible pneumonia, possible cancer (due to a "shadow" on the lung x-ray), and/or being too fat to breathe properly. Finally, a triage nurse pointed to a possible pulmonary embolism (PE). A PE is a known risk factor of major joint surgery on the leg or hip. After treatment of the PE, which included a one week hospital stay, my breathing improved... only to get worse again after 2 months or so. An echocardiogram, which was done as a prerequisite for a respirologist referral, gave me the diagnosis of secondary Chronic Thromboembolic Pulmonary Hypertension (CTEPH).

Only about 1% of joint surgery patients develop a PE, and only 4% of PE patients will develop CTEPH within 2 years. It is estimated that CTEPH occurs in about 8 to 40 cases per million. I developed CTEPH, but funnily enough, I can't seem to buy a winning lottery ticket.

I remember being in my GP's office, where I was told that the results of a recent echocardiogram showed that I have pulmonary hypertension. It was decided that I should see a respirologist sooner than the previously scheduled appointment in upcoming September. All I heard was hypertension: surely there was a pill or something that would "fix" this? Since there was no urgency in my physician's tone of voice, I did not feel much apprehension about the diagnosis. This was until I met with the respirologist, who told me that the average life expectancy of untreated CTEPH is about 3 years. It felt like the floor was falling out from under me. The respirologist explained the "plan of attack" in detail, from possible surgery to treatment with drugs. At that time, a drug specifically for CTEPH was not available, but one was expected to become accessible by the end of the summer. Since November 2013, I've been on Adempas, which is the only medication available in Canada specifically for CTEPH.

"The respirologist told me that the average life expectancy of untreated CTEPH is about 3 years. It felt like the floor was falling out from under me."



Can you believe the misdiagnoses that Juergen faced?

The most memorable moment of my PH journey occurred very early on, shortly after diagnosis. At the first meeting of the London PH support group that I attended, Grace Wickenheiser introduced herself and said that she had been diagnosed in 1995 and had been living with PH for over 19 years. Up to that point, my brain had been spinning around the 3 year survival statistics, and everything I had read which indicated that a survival of eight years or longer was considered "long term". To witness people like Grace who were thriving with PH gave me a new lease on life with my disease.

I also realized that, relative to others, I was diagnosed early: only 6 months from my initial breathing problem. This was because my PE was found and I was seen and treated by a respirologist. Those 6 months were a long and scary time of not knowing why I could not breathe properly. However, I'm grateful that it was a short delay compared to many others' experiences.

For me, the hardest part of living with CTEPH was tolerating my newfound limitations. As a productive member of the workforce, I found it the hardest to accept the fact that I would not be returning to work, and would have to take an early disability retirement. However, it's been very inspiring to me to participate in meetings of the PH Support Group in London and meet other people living with PH. Also, participating in various Facebook support groups is motivating because it allows me to see how other PH'ers deal with the disease.

I'm also grateful for the support of my immediate family: two big thumbs up to them. My wife and two daughters are very supportive, sometimes maybe a little too supportive, but it is very much appreciated. Most of my acquaintances do not realize how life threatening PH is, because I do not look sick. This makes my family's love and care all the more valuable.

To those who are newly diagnosed: try to accept the disease for what it is. Though it's not curable yet, it is treatable. Have full trust in the treating specialists and make the lifestyle changes that are needed to survive PH.

Contributed by: Juergen Buettemeyer, PH patient, London, ON.

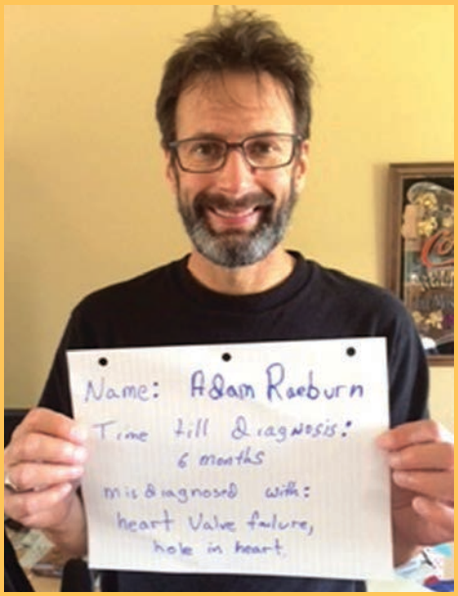
"Most of my acquaintances do not realize how life threatening PH is, because I do not look sick. This makes my family's love and care all the more valuable."

From "I must be out of shape" to Flolan

It started in May 2005 when I was having trouble mowing the lawn: the lot is only 100 feet square and it was taking me 2 hours to mow. In July, I went to the ER when I thought that I was having a heart attack. They did a chest x-ray EKG and checked my vitals, which were all normal, but they did notice that my heart was enlarged a bit. While in the ER, the doctor kept asking if I had a burning in my chest. I did and I tried to explain how it felt. It wasn't like heartburn, but more the feeling of just having run around the track. My lungs felt like they were screaming for air. I was referred to a heart specialist in Saint John, who checked for a misplaced heart valve or hole in my heart. When these were ruled out, I was sent to a lung specialist.

These appointments took all summer and we still had no idea what was wrong with me. I still felt like someone standing on my chest. The doctor suggested that the night shift I had been working for the last 4 years might be catching up with me. When he suggested I try and get out and walk, my kids complained that I went too slow. A 3-block walk took 20 minutes one-way.

Sept 30th was the last day I went to work. I was coughing so much that I was not able to take more than one call (I work in a call center) during the shift. I felt weak, short of breath, tired and wasn't able to concentrate. When I visited the ER again feeling that shortness of breath, the EKG and all vitals were a little high, but not alarming for an out of shape 39 year old. I was sent home to wait and see the lung specialist that I had an appointment with in 2 weeks' time. Two days later, I went back to the ER and told the intern that if I was sent home again, I felt like I'd be dead within 2 weeks. I was admitted before lunch. Over the next 10 days, the doctors completed a series of tests, all of which I was scheduled for up to six months later because they could not get me in sooner. As Sherlock Holmes said, "Eliminate the impossible and whatever is left, however improbable, must be the truth." And I was diagnosed with PH. The Saint John lung specialist told me there was not a lot that could be done here for me, and that I should get to a clinic in London, ON. At this point I really did not have a lot of questions, as it really had not set in as to the complexity of the disease.



Adam shared his PH story for PHA Canada's Early Diagnosis facebook photo campaign in November 2014.

During a morning newscast on a local radio station, my wife Cheryl heard a report on a child that had the same disease, and who was trying to raise awareness. She called the radio station and was able to get the number of the mother and called right away. This mom was more than happy to bring us information on PH, and dropped off a few flyers and a large book called PH: A Patient's Survival Guide, which has been a great resource for myself and my family.

By early December, I was feeling weaker and had been retaining fluid since I came home from the hospital.

I live in a 4 level split home and was going up and down the stairs all the time. My family doctor advised that I find a level that would be best for me and stay on it. I chose the family room and started living on the pull out couch. On January 11th, I met with the doctor in London, where they determined that my condition had worsened in the past 8 weeks. They estimated that I had 6 to 8 months to live, and if treatment did not work, there would not enough time to try anything else.

On January 18th, I was started on Flolan. For those that are on Flolan, you know how intense the next 3 weeks are and what you need to know to become one with your pump. I have 2 girls, ages 6 and 10. I have therefore named the pump and his name is Ed. This I think helps the girls to understand that this is something that daddy will have with him all the time.

Our girls had stayed with friends and family while we were in Moncton (it is an hour and half drive from were I live). The first time they came up to visit, I was feeling and walking so much better than I had when they last saw me at home. They kept saying "Daddy, slow down, you're going too fast." This was the greatest thing I had ever heard.

Since my diagnosis, we decided it was best to move and built a single level home in a new subdivision. We made sure the house would accommodate a wheel chair if we ever should require one. The house is a bit smaller and has an open concept. In addition, after facing several line infections 3 years ago, I was switched from Flolan to Remodulin. While I do have to deal with some site pain, Remodulin does not need to be kept cold. It has a 30-day life span in the vial, and up to 4 hours in the body. The pump is 1/3 the size of the pump used with Flolan, so all of this makes life a little easier. Since having this disease, the most inspiring part of life is watching my children grow up, and seeing how each day without sickness is a blessing. I look forward to as many more years as possible with my family.

Since having this disease, the most inspiring part of life is watching my children grow up, and seeing how each day without sickness is a blessing.

Contributed by: Adam Raeburn, PH patient, Saint John, NB.

Ten years and counting

LIFE WITH PULMONARY HYPERTENSION

I was diagnosed with pulmonary hypertension about 11 years ago, when I was 20. This final diagnosis took place after I'd seen 2 or 3 specialists in Poland and had been diagnosed with pulmonary fibrosis, and after seeing another specialist in Canada. I initially felt relieved that I was officially diagnosed and there were treatment options available. However, it was also frightening to find out the disease is progressive and potentially fatal. I still assumed things wouldn't be much different going forward, since I had been experiencing symptoms for a long time. Unfortunately, life changed more than I expected.

> ***"It was surprising to see that so many people were affected by the disease, but also comforting to know I wasn't alone."***

One of the most memorable moments I experienced was when I was told that I would need to go on Flolan. That's when I realized how serious the disease was. The hardest part about living with PH is not being able to do all of the things I want to do, for example, playing sports and other activities. Also, being on Flolan - despite it being very effective - is a major inconvenience.

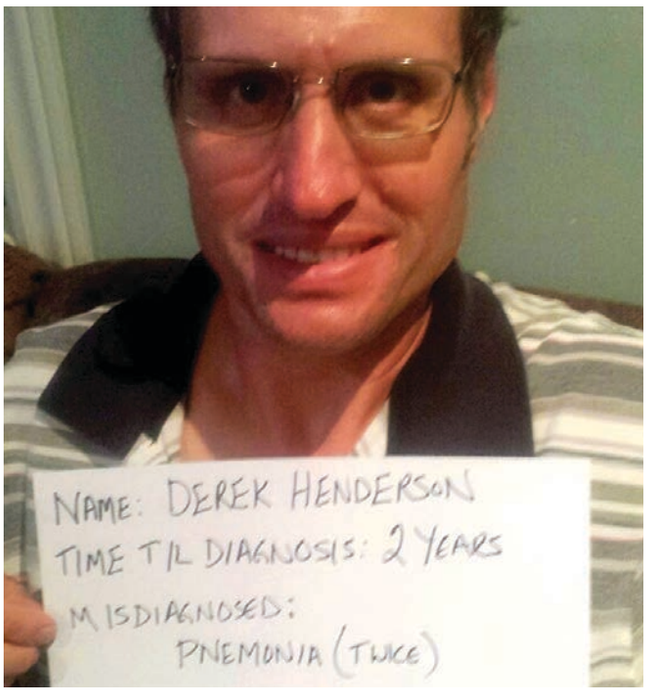
The most inspiring part of life with PH has been learning about the amount of research that is taking place around the different forms of this illness, as well as meeting all the people in our PH support group. It was surprising to see that so many people were affected by the disease, but also comforting to know I wasn't alone. My family and friends have also been very supportive.

If I could offer my advice to anyone who has only recently been diagnosed with PH, I would tell them that living with PH can be very frustrating because of all of the limitations that you now have. At the same time, there is a lot of hope, and it's important to focus on that.

Contributed by: Marcin Gozdzik, PH patient, Edmonton, AB.

Marcin doesn't let pulmonary hypertension slow him down, and continues to live life to the fullest!

Snorkeling after CTEPH: the story of my life-changing surgery



Derek sharing his CTEPH story and misdiagnoses for our Early Diagnosis facebook photo campaign.

For me, everything started in 2010. My wife was going through cancer treatment, and I ended up being short of breath for no real reason. I didn't have a cold or anything. It gradually got worse to the point where I went to a clinic and was eventually diagnosed with pneumonia. The doctors told me that I was probably just run down from the stress of dealing with my wife's cancer. But 6 weeks later, my symptoms returned. Doctors suspected pneumonia again, but an initial x-ray came up clear. However, after a second look, they did find pneumonia. I was treated and it seemed to go away.

Fast forward 2 years. My wife always says it started with a hill. I bike a lot: about 150k per week in the warm weather. In early June, I suddenly could not make it up a small hill. I went to the clinic again, but they could find nothing wrong. Summer ended, fall started and I found I was getting short of breath doing less and less. What would have been a 45 minute bike ride was now taking 2 hours. I went to doctors and clinics a few times, but they couldn't find anything. They scheduled some pulmonary and cardiac tests, but because I was not "urgent", they were months in the future. Only a few weeks later in late October 2012, I could no longer make it up a flight of stairs without being short of breath, so I went to the hospital. There, I was admitted into an ER and was finally diagnosed with chronic thromboembolic pulmonary hypertension (CTEPH). I had seen about 6 specialists before this diagnosis was made.

I remember the initial relief that I had finally been diagnosed with something and was getting treatment, because for the months before that I had been getting worse and worse and no one had been able to tell me what was wrong. However, I was now concerned that I wouldn't be able to be as physically active as I once was. I also live in a house with a lot of stairs, so I thought we might have to move... and of course I was worried about what the future would hold.

One of the challenges in living with CTEPH is that most people do not know what it is or what it's like. A lot of people did not believe I was as sick as I was, because I looked healthy. Close friends are usually quite supportive. One employer was not supportive and more or less dismissed it.

Another hurdle presented itself in 2013 when I received an email which confirmed that I would be undergoing pulmonary endarterectomy (PEA) surgery. I was given only 6 days warning between the time I was told I was going to have the surgery, and date the procedure would take place. This was an added challenge because I hadn't yet warned anyone that I was even being considered for the surgery, much less told anyone what it would involve. PEA is a traumatic open heart surgery, where you are in effect dead for a period of time while doctors remove your blood clots. Going through that was probably the hardest part of my CTEPH experience, because I was also thinking about what my family was going through and all of their worry for me.

"About 18 months after my initial diagnosis, I went on the trip of a lifetime to Bonaire, a Caribbean island, to snorkel for 10 days. I was able to do as much as I wanted to when I was there, which was incredible."

The PEA surgery was successful for me: I'm now able to be more or less fully active again. This is definitely inspiring. About 18 months after my initial diagnosis and only 6 months post PEA surgery, I went on the trip of a lifetime to Bonaire, a Caribbean island, to snorkel for 10 days. I was able to do as much as I wanted to when I was there, which was incredible.

My advice for those being diagnosed with PH is to not be afraid to ask for help. There will be bad days at times, but they will pass. Speak with fellow survivors: I remember speaking with another patient who was helpful with encouraging me to get treatment. When someone has gone through it, and is confident that you can too, it's very motivating. Also, make sure that your family understands what you're dealing with: it will help both you and them.

Contributed by: Derek Henderson, PH patient, Toronto, ON.

Ask a Nurse:

How your family doctor can help you manage your PH

I was employed as a nurse for twelve years before I encountered a patient with pulmonary hypertension. As pulmonary hypertension is a rare disease, many physicians in general practice will only come across one or two patients with pulmonary hypertension in their whole careers. These patients are often sent to expert centers to be diagnosed and treated before returning to their local communities.

Even though your family doctor may not be familiar with pulmonary hypertension, they can be a great resource for you. It is important for you to maintain a good relationship with your family doctor as only one third of patients in BC live within an hour drive of a PH center. Patients across Canada experience similar geographic challenges. Your relationship with your family doctor is important for other reasons as well.

Nurse Lisa (second from left) at Vancouver General Hospital.

Your family doctor can help you:

- Manage and treat the other medical conditions you may have besides pulmonary hypertension. Pulmonary hypertension clinics do not have enough resources to manage your blood pressure, diabetes, or infections and your family physician is the best resource person for those concerns.

- Identify and treat other common medical conditions that can be causing your shortness of breath. Not all shortness of breath can be attributed to pulmonary hypertension, especially if your pulmonary hypertension is mild or well controlled with medication. Other common conditions that can cause shortness of breath include coronary artery disease (poor blood flow to the heart caused by a build up of plaque in the arteries in the heart), atrial fibrillation (irregular beating of the heart), chronic obstructive pulmonary disease (damage to the lungs most often caused by smoking), or pneumonia (lung infection).

- Assist with monitoring and treating heart failure. If you live far from a PH centre, it can be difficult for your PH care providers to manage your heart failure. Your family physician can help by checking your kidney function and electrolytes (especially potassium, which is affected by most diuretics) and by increasing or decreasing your diuretics depending on your weight, your symptoms and your blood work results.

- Help you monitor your own condition. Sometimes it is difficult on a day to day basis to assess your symptoms and tell if you are better or worse. If you see your family physician regularly, they can help track how your breathing is and how you are overall functioning. Your physician may be able to identify early warning signs that you need to be evaluated by the PH clinic.

- Ensure your vaccinations are up to date. All patients with pulmonary hypertension should have a yearly vaccine for influenza and one time pneumococcal vaccine to protect against 23 types of streptococcal pneumonia (sometimes a second dose is needed).

- Access resources in your local community. Your family doctor may be able to tell you about valuable resources in your community that can help you maintain your health and well being such as pulmonary rehabilitation, support groups, social workers, counselors, and dieticians.

You can also help your family doctor take care of you by:

- Valuing their time. Family physicians are paid differently than specialists and they may have less time to spend with you. Focus on the most important question you have and have your thoughts in order. You may need to schedule a follow up visit if you have more than one concern.

- Keeping your family physician up to date on what is happening with your health. Ensure they are receiving information from your visits to specialists and have copies of the tests results for any investigations being done.

- Keeping an up to date list of medications so that your family physician can check for interactions if a new medication is needed. Most medications your family physician will prescribe will not interact with your PH medications, but some will. If there are any questions, check with your pharmacist or your PH care provider.

- Having the phone and fax number of the PH clinic available so it is easy for your family physician to call the clinic if it is needed.

Contributed by: Lisa Lee, Nurse Practitioner, PH Clinic, Vancouver General Hospital

Dr. Mielniczuk co-chairing a session at PHA Canada's 2013 National PH Conference.

Meet your medical professional:

Dr. Lisa Mielniczuk

Dr. Mielniczuk is a PH-treating physician and the Medical Director of the PH clinic in Ottawa, where she has worked since 2008. She is also a member of PHA Canada's Medical Advisory Committee, offering expertise on PH-related publications and campaigns, and has led multiple session at PHA Canada Conferences. We are so grateful for Dr. Mielniczuk to share her journey and passion for the PH community with us in this e-interview for Connections magazine.

Where did you begin your medical career, and in what field?

I completed medical school at McMaster University, and did a residency in Internal Medicine at Queens. I then went on to do a cardiology fellowship in Ottawa and did a fellowship in advanced heart disease (heart failure and transplant) at Harvard University/Brigham and Women's Hospital in Boston, Massachusetts. During this time, I also completed a Master's Degree in clinical epidemiology at the Harvard School of Public Health. After completing my training, I started working at the University of Ottawa Heart Institute where I am today.

How did you first learn about PH? What drew you towards the field of PH: why did it interest you?

I was exposed to PH as a cardiology resident. We had a young patient with a new diagnosis of severe PAH – she was very ill and required critical care. Fortunately we were able to stabilize her, however at the time there was no PH clinic in Ottawa, and she had to be transferred out to be offered further care and management.

I am a heart failure doctor, so spend much of my time working with very ill patients who have advanced disease. I consider it a privilege to work in the field of PH. These patients have tremendous stories, and the journey they take is remarkable. It is wonderful to be part of that journey and to positively affect their lives and the lives of their families in such a significant way.

How long have you worked in a health-related field, and how long have you worked with individuals affected by PH?

I have been the Medical Director of the PH clinic in Ottawa since I started on staff here in 2008.

How did you become involved with PHA Canada?

I became involved with PHA Canada through engaging with patients, and working with patients, families and colleagues across the country. I've also attended and presented at several of PHA Canada's National PH Conferences.

What do you enjoy the most about your work as a PH Specialist?

I am honoured to be part of this team of incredible health care providers, all of which share a common vision and passion for improving the lives of patients with PH.

What do you find most frustrating?

It's very difficult to deal with issues of access and limitations to medical therapy for our patients.

What has been the most inspiring part?

Without a doubt, watching patients grow, heal and improve. Witnessing the resolve and determination that patients have to live every day to the fullest.

What advances have you seen in the treatment of PH patients since you started practicing? What would you find to be the most encouraging advances currently taking place?

I think that this is one of the most exciting fields in medicine right now. We have come so far in such a short time. There are new therapies available, and others on the horizon. We are learning more about this disease every day.

As you know, early diagnosis is an important strategic area for PHA Canada. Why do you feel that the issue of early diagnosis is this critical for patients? What steps can we take to ensure an earlier diagnosis for those affected by PH?

I would encourage any person who may have some of the symptoms of PH to see their doctor and discuss their feelings. Any patient who has a risk factor for PH should also discuss with their doctor about the possibility of PH. It is critical that we diagnose this disease as soon as possible, as we know our ability to improve symptoms and minimize risk is greatly enhanced if we can pick up cases before patients become very ill.

"I consider it a privilege to work in the field of PH. These patients have tremendous stories, and the journey they take is remarkable. It is wonderful to be part of that journey and to positively affect their lives and the lives of their families in such a significant way."

Can you recall one specific memorable moment or conversation with someone affected by PH that was particularly meaningful to you? Can you describe it?

I have a patient who had PAH who was very ill, and her disease progressed despite everything we could offer her. She was very reluctant to consider transplant as an option, but eventually realized that this was her best option. She had a very long recovery and we were very worried that she might not get through. She amazed everyone – despite all the odds, she pulled through and her resolve and will to fight was stronger than anyone I have ever met. I am amazed at her courage and strength, and I still think about it often. I consider myself very fortunate to have been involved in her care.

If you could sum up everything you've learned about PH into a couple of sentences that you would want to share with someone who is newly diagnosed or newly affected by PH, what would those be?

This is a serious disease. There will be ups and downs. But the options for treatment are growing every day, and these medications can substantially improve your sense of well being and wellness. You are supported by a team of health care providers who will stand by you and navigate this with you and your family. You are not alone.

Contributed by: Dr. Lisa Mielniczuk, BSc, MSc, MD, FRCPC, Ottawa, ON

Arjun Pandey is a Grade 10 student and one of the youngest PH researchers we know. Arjun conducted research on the effects of obesity, weight loss and lifestyle modification on pulmonary hypertension. His study took place at the Cambridge Cardiac Care Centre, and he presented his findings at the Canadian Cardiovascular Congress 2014. Arjun is the youngest person to ever present at the Canadian Cardiovascular Congress, and has also become a PHA Canada knowledge philanthropist and will be continuing to share his research with us. We're grateful to Arjun for continuing to investigate into pulmonary hypertension and look forward to seeing what he achieves next.

Effects of obesity and lifestyle interventions on mild to moderate pulmonary arterial hypertension

While many dietary strategies exist for individuals with elevated systemic blood pressure (blood pressure in the non-pulmonary arteries), limited studies have been done on the effects of dietary changes and lifestyle interventions in pulmonary arterial hypertension. I conducted a study recently as part of my high school science fair project with the assistance of Dr. Sanjay Mehta to evaluate the impact of obesity as well as the effectiveness of the DASH (Dietary Approaches to Stop Hypertension) diet to reduce pulmonary pressures.

The study consisted of a test population of 60 people, 30 with mild to moderate PAH and 30 people with normal pulmonary pressures. Participants had their height, weight, and pulmonary pressures assessed by echocardiography before and after a 2 month dietary intervention period wherein patients were prescribed the DASH diet. The DASH diet is recommended by the American Heart Association to control systemic blood pressures but has never before been assessed in patients with pulmonary hypertension. The DASH diet is rich in fruits, vegetables, fish, low-fat dairy products, lean meat products, and emphasizes a low sodium intake.

At baseline, before any changes in lifestyle were initiated, a very strong correlation was observed between the Body Mass Index (a commonly used measurement system for obesity) and the pulmonary pressures of the individuals. After two months of dietary changes, PAH participants who achieved a weight loss of 5% or more, while on the DASH diet, had an average 25.3% reduction in pulmonary pressures (11.3 mm Hg systolic reduction). Those who achieved a weight loss of 1-5% while on the DASH diet had a slight reduction in pulmonary pressures. On the other hand, those who did not adhere to the diet and whose weight increased over the two months had an average increase in pulmonary pressures.

The results of this study may imply that in individuals with mild to moderate pulmonary hypertension, effective weight loss strategies and lifestyle changes (such as the DASH diet), may be a strategy to consider, in discussion with their health care providers. This is a preliminary study in a small group of individuals; lifestyle interventions warrant further investigation in this condition. This study was limited to individuals with mild to moderate pulmonary hypertension and its results cannot be generalized to individuals with more severe or advanced conditions. Always consult with your health care providers before making any changes.

Contributed by: Arjun Pandey, PH researcher, Waterloo, 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.

Why do we still not have a cure for PH? And other research questions

In this issue of Research Corner, I will provide you with some insight into the challenges facing PH research, as well as answers to some other questions.

Why do we still not have a cure for PH?

In order to be able to look at curing a disease, scientists have to know why and how the disease occurs and how it progresses, which requires understanding which systems, organs and cells are involved in the generation of the disease. This knowledge allows the discovery of ways to stop and ultimately reverse that progression of the disease. In PH, there are some challenges we face with understanding how and why PH develops, and that limits our ability to develop a successful cure.

The first is the origin of PH: PH is classified into five groups and each has its own subgroups. On the clinical level, patients can appear similar, displaying increased pulmonary artery pressure, increased pulmonary vascular resistance and right ventricle thickening. However, each subgroup of PH patients is unique at the cellular and molecular level, due to different causes of the disease. For example, group II PH is due to left heart disease, while group IV is due to blood clots in the lungs; both result in PH, but need to be treated differently. This uniqueness dictates which therapies will work while others will not, making it difficult to find a common cure that works for all these conditions.

The second issue is progression of PH: PH in the early stages is almost asymptomatic, and many PH patients present to the clinic at the later stages of disease. At these stages, the lung blood vessels display a more complex pathology, including the blockage or closing of blood vessels and complex lesions, as opposed to the earlier stages where the blood vessels are only narrow. This makes it more challenging to reverse the condition.

The third issue is the "multifactorial" nature of PH. Several cell types are involved in PH disease pathology, which can differ depending on the origin of the disease. Endothelial, smooth muscle, and immune cells can all contribute to PH pathology and progression. This is one of the major complications for finding a cure, since some drugs can only work on one cell type but have opposite effects on another, which means that they fail in producing a significant improvement in patients.

On the horizon: looking at the above, you might get a bleak image of PH research, but you should not. The field of PH therapeutics is evolving very rapidly and current approved therapies have significantly improved patient life and slowed down disease progression. Scientists are, and will always be, working hard to discover new pathways involved in PH pathology, and develop novel therapeutics to find a cure for this devastating condition.

Why do some PH patients have a drop in their peak VO2, while others do not?

Peak VO2 is a measure of the maximum rate of oxygen consumption. Variability in peak VO2 occurs because VO2 doesn't only depend on PH progression: it's also affected by underlying other cardio or pulmonary diseases; depression or anxiety. This explains the variability.

What is the difference between a CCBs (Calcium Channel Blockers) true responder and a non-responder?

Calcium channels are the major channels regulating calcium uptake into the cell. Calcium is important in PH because it can control the contraction of smooth muscle cells, thus blocking calcium intake, using CCBs, can result in more relaxed blood vessels in the lungs. To determine if CCBs can be used, an initial acute vaso-reactivity test using a vasodilator factors (such as nitric oxide inhalation) has to be performed. If the pulmonary pressure decreases with this acute exposure, then the patient can be treated with CCBs.

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 & Molecular Medicine, Faculty of Medicine, University of Ottawa

My experience in cardiac rehab

Jill staying strong and healthy!

One of the challenges we face as PH patients is how to stay in shape. What is enough exercise? What is too much? How can we begin safely? A referral from the PH Clinic to a cardiac rehab program at my local hospital (Royal Jubilee in Victoria, BC) gave me the answers.

The cost of the program was just under $400 for three months, and included two 90 minute sessions per week. Our province of BC does not cover this cost, but it can be claimed as a medical expense on your income tax. You are seen by a cardiologist first, then attend an introduction workshop, and then choose whether to sign up at the hospital, a community centre, or with a private trainer. The workshop covered the functioning of the heart and types, symptoms and causes of heart disease. We learned about modifiable and non-modifiable risks and how to improve heart health through exercise, diet, stress reduction and addressing depression, which is a common symptom of those with heart disease or chronic illness. I was so impressed with the multidisciplinary approach: it wasn't just an exercise program; it also provided opportunities to meet with a nutritionist, social worker and psychiatrist. I cannot begin to list all the valuable information I obtained from these consults: I needed a binder just to hold all of the materials and printouts!

When I first stepped into the gym, I noticed it was lovely and bright, with an array of exercise equipment. Music was playing and the participants began and ended each session by checking their blood pressure, weight, and blood sugar (for those with diabetes). We had an individualized session with the lead physiotherapist, who tailored a program for each of us to follow each day. I had a sheet which told me which machines to use, as well as which weights, amounts of time, and repetitions. Sticky notes were available, which I attached to everything to help me remember. Each session started with a slow warm up on a recumbent bike. The cardiac patients were allowed to use both arms and legs at the same time, but as a PH patient, my program instructed me to alternate between the use of one and the other.

I received careful instruction on how to use a variety of bikes, treadmills, free weights, rowing machines and presses for shoulders, legs, triceps and lats. Since my balance was also an issue for me, I had some balance exercises to be done on a thick mat in front of a mirror.

The gym had multiple colourful posters to remind you to breathe in "The Safe Zone". The warm up kept you breathing in the level of 0-2, which meant exercise easy enough to complete while singing. The moderate exercise was your targeted "Green Zone", 3-5: an exercise level where you'd have enough breath to talk. If you moved into the "Red Zone" of 6-10, it meant you were gasping and either needed to rest or slow down.

The physio watched me closely enough to notice whether I was breathing with flared nostrils or pursed lips. These are behaviors that even I was unaware that I had developed, which we PH patients do in an attempt to increase our intake of oxygen. They meant it was time to slow down. Other PHers know very well that there are good days and there are bad days. I found that my PH was very good at drawing a line in the sand: it let me know with dizziness or increased shortness of breath that I was clearly working too hard and over-exerting myself. Both the staff and I learned to alter my program until excess edema was under control and I could feel the benefits of increased activity. By the end of the three months, I had increased my strength, stamina and had a statistically significant improvement in my six minute walk test. These were fantastic results which I was very happy with.

The psychiatric consult provided me with tools for stress reduction. The nutritionist helped me begin a safe program of weight loss and promoted better eating habits. I learned how diet affects anti-coagulants, and was given information on supplements and wheat and dairy sensitivities. Soon I begin a series of workshops called "Craving Change", which were about our relationship with food and understanding why we eat the way we do. The Social Worker made sure I knew what services were available in the community as the need arose.

Most importantly, I left my cardiac rehab program with information on how to continue with this healthier lifestyle and how to replicate it at home, even using the exercises I had learned. While it may seem a daunting task, it just takes one step at a time. Start slowly and know that some days you will need to rest. In the long run, the effort will make such a difference!

Contributed by Jill Morton, PH patient, Victoria, BC.

I've already been to a PH Conference, so why should I attend?

If you have attended one of PHA Canada's National PH Conferences in the past, you may feel like there isn't any reason to go again. You have been to the sessions, you're aware of the current research going on in pulmonary hypertension, and you have already connected with wonderful PHriends with whom you share your experiences. So what it the point of attending another Conference? The truth is that every Conference is different, and this one may be the most different yet. Here are a few reasons to register for our 4th National PH Conference today.

You've never been to sessions like these.

Some of the regular sessions, focusing on PH research and resources for living with PH are essential to those who may be newly diagnosed. However, that doesn't mean that all of the sessions are the same as those you may have experienced before. At our 2015 National PH Conference, one session will feature a live cooking demonstration, where an experienced cook and certified instructor will cook a low-sodium dish from beginning to end. Another will help participants to develop a personal toolkit for coping with stressful situations. The presentation will include information on using breathing techniques and various forms of meditation as a means of stress relief. The emphasis for the sessions at this Conference goes beyond aiming for physical health with PH: our hope is to provide patients and caregivers with information on navigating relationships, taking charge of emotional wellness and effective communication for both social ventures and advocacy. Even if you've been to every Conference before, we believe that at this Conference, you will learn something new.

You might meet someone who could change your entire perspective.

Many PH patients and caregivers already have a wonderful support network and close friends that they lean on for life's difficult moments. That being said, no two PHriends are alike. At our 4th National PH Conference in Montreal, even veteran Conference attendees are guaranteed to meet someone new. It may be someone who is newly diagnosed and has a different perspective on the PHight. It may be someone who has lived with PH for longer than you thought was possible. Conference provides the opportunity to connect with individuals who live the same reality as you do, but they might see life in a different light. These connections are priceless, and last for long after the event is over.

Or, someone else may be inspired by YOUR story.

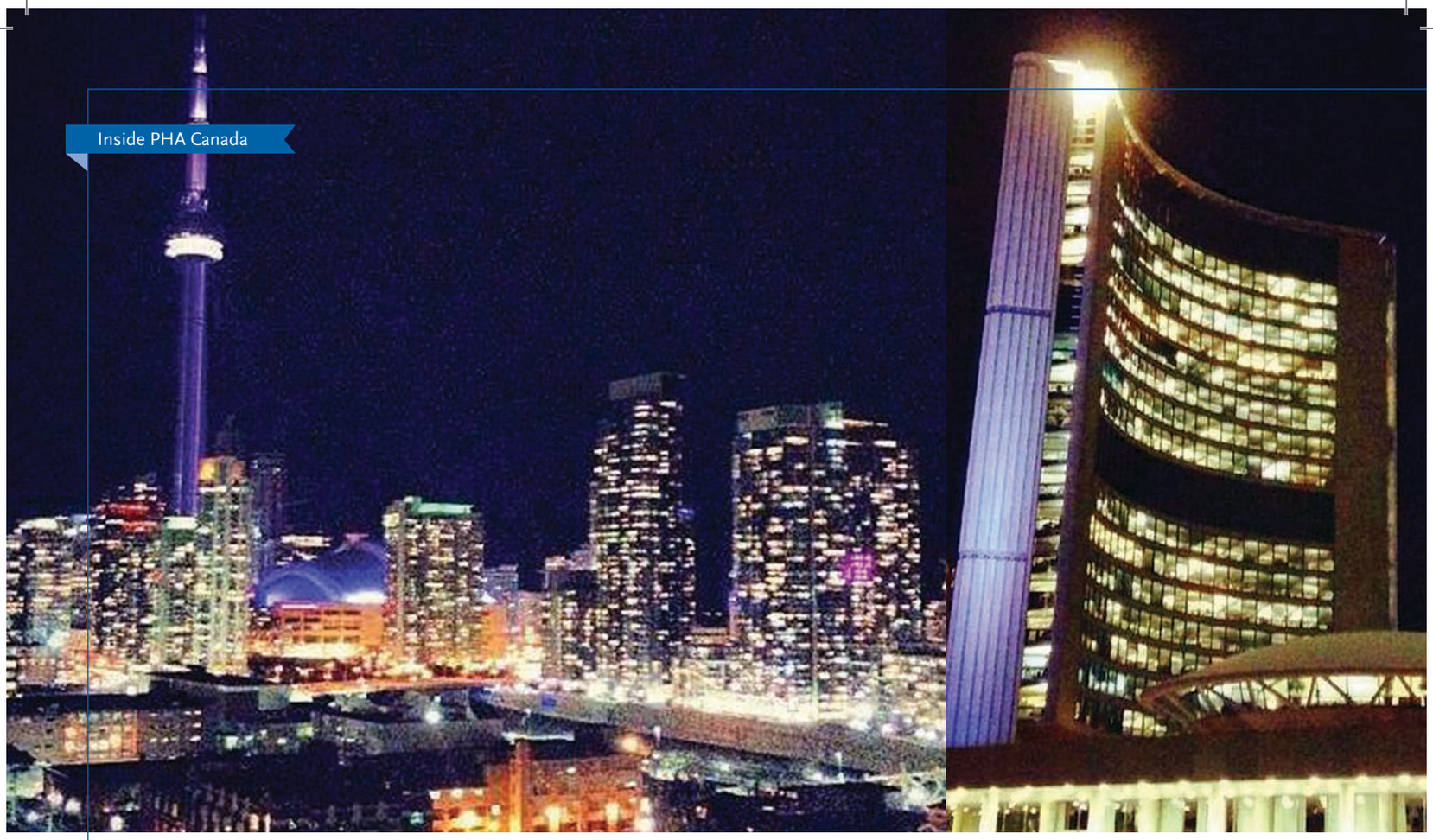
You may feel as though you have everything that you need for your PH journey. If this is you, you hold more power than you know. To be content and stable in your life with PH is an incredible position from which you may become a true inspiration to others. If you are a PH patient who has it all, please consider that people at Conference may need YOU more than you need them. Newly diagnosed patients who come to Conference take significant strength from witnessing people like you who are at ease with their PH. Know that you may make an enormous difference to someone's life just by sharing your story.

Conference makes change possible by bringing us together.

PHA Canada's Conferences are the single largest gatherings of PH patients in Canada. This critical mass of people who care can make an extraordinary impact in advocacy and in media, because Conference gatherings are where ideas are shared and plans are made. An amazing amount of energy and togetherness arises out of Conference, and it fuels the PHight for the year ahead. Helen Keller said "Alone we can do so little; together we can do so much", and it could not be more true.

We hope that if you weren't sure if you needed to attend another Conference, that you will still join us at the 4th National PH Conference in Montreal from May 1-3. We cannot wait to see what we will all create together, and we can't do it without you. Visit phacanada.ca/2015conference to learn more and register.

Contributed by: Bronwyn McBride, Communications Associate, PHA Canada



Paint Canada Purple for World PH Day: What YOU can do

World PH Day is on May 5th, 2015. Inspired by an incredible initiative by Loretta Chu in 2014, our primary campaign this May will be 'Paint Canada Purple'! For this campaign, we are requesting PH advocates across Canada to request that their local monuments be illuminated in periwinkle for World PH Day on May 5th. May is just around the corner, and now is the perfect time to begin approaching the offices of your local monuments to request that they be lit up.

Last year, PH patient and long-time supporter Loretta Chu single-handedly succeeded in having the CN tower AND City Hall in Toronto illuminated in periwinkle to raise awareness of PH on World PH Day. Her success inspired Carolyn Doyle-Cox to achieve illumination of Niagara Fallsand Jennifer Merritt requested the Fort Erie Peace Bridge, and BC Place was illuminated after they received information about World PH Day. This year, Loretta has gone above and beyond and developed this list of monuments which do illumination by request for different causes. They are:

BC Place, Vancouver, BC
Science World, Vancouver, BC
Jack Poole Plaza, Vancouver, BC
Clock Tower, Victoria, BC
Highlevel Bridge, Edmonton, AB – ACHIEVED by Lynn-Marie Cox
Olympic Plaza, Calgary, AB
Calgary Tower, Calgary, AB
Telus Spark Science Centre, Calgary, AB
City Hall, Airdrie, AB
Town Hall, Okotoks, AB
Tiff Bell Lightbox, Toronto, ON
RBC Zipper, Toronto, ON
City Hall, Toronto, ON
CN Tower, Toronto, ON – Requested by Loretta Chu
Niagara Falls, Niagara, ON
Peace Bridge, Fort Erie, ON
Heritage Building, Ottawa, ON
Maestro SVP, Montréal, QC
Legislative Building, Fredricton, NB
City Hall, Fredericton, NB
Gazebo in Wilmot Park, Fredericton, NB
Cabot Tower, St. John's, NL
City Hall, Halifax, NS

Monuments across Canada alight in purple in 2014: how many more will we see this year?

While all of these monuments are open to being illuminated for special causes, this is by no means a comprehensive list of all potential monuments across Canada. A simple request to the civic authorities in your town or city could result in lighting up many other buildings! We need YOUR help in order to truly Paint Canada Purple in every province.

TO CONTRIBUTE TO THIS CAMPAIGN:

1. ***Think about which monument YOU would like to see lit in periwinkle. Once you've decided, seek out the email address or mailing address of someone connected to the monument who could help with your request.***

2. ***Download our easy template letter from phacanada.ca/paintcanadapurple. This letter is a general request to illuminate a monument, and it will only take you moments to customize it to your monument and your area!***

3. ***Send your letter or email and wait to hear back. Don't be afraid to send several requests if you don't get a reply!***

4. ***Make sure to let us at PHA Canada know about the monuments that will be illuminated. This will help us to get critical media coverage, which in turn will exponentially increase awareness of PH. The more monuments lit in purple across Canada, the more likely that local and national media outlets will run a feature on them. So make sure to let us know about your successes!***

TIPS:

Multiple requests?

You might be thinking "What if some of these authorities get several requests for illumination, because others from the PH community are writing to them too?" In our experience, when MORE individuals want something to happen, the more likely it is that the authorities take notice. So don't worry if you think the monument has already received a request: feel free to ask anyway!

Do smaller monuments count too?

Absolutely! Even if you live in a small town with a little city hall, every single monument in periwinkle will promote awareness on World PH Day.

We are imagining all of Canada lit up in purple for World PH Day, and the incredible potential for increased PH awareness. Please join us in Painting Canada Purple for May 5th, World PH Day! If you have any questions for this campaign, please email bmcbride@phacanada.ca for support.

Contributed by: Loretta Chu, PH patient, and Bronwyn McBride, Communications Associate, PHA Canada

Choose happiness, despite life's challenges: a report on the 2014 HTAPQ conference

Pulmonary hypertension is a serious disease caused by high blood pressure in the arteries of the lungs. People diagnosed with PH experience a marked decrease in their quality of life, and despite medications and treatments, they still suffer from symptoms like breathlessness, palpitations, dizziness, swelling in the legs and feet, and more. Most PH patients are unable to work, so their incomes are dramatically reduced. Finally, PH patients often experience isolation because the disease is so rare.

In 2006, Mr. Denis Cormier from Plessisville (whose daughter had been diagnosed with PH) decided to found an association to bring together those who were suffering from this chronic disease. It was then that the HTAPQ (PH Association of Quebec) was born. The Association provides financial support to PH patients that need it. Another important mission of ours is to create opportunities for PH patients to come together and meet one another, so that they may be able to discuss day to day life with PH and share their experiences with others who can truly understand.

To that end, every two years, the Association organizes a large scale conference for all PH patients in Quebec, as well as their caregivers. In 2014, the 4th provincial conference was held in Longueil, QC from September 19th to 21st. The theme of the conference was 'Choose happiness, despite life's challenges'.

This amazing opportunity allowed PH patients and caregivers to socialize, share and become more informed about their illness. Beyond addressing physical challenges, the goal of the conference was to provide tools to patients and caregivers in order to minimize the social and psychological consequences of living with PH. This year's conference session topics were chosen based on the findings of a recent national survey (PHA Canada's Burden of Illness Survey) and through the support of our PH-treating physicians.

One of the sessions, presented by Marie-Chantal Brisson, was titled 'Learning how to eliminate stress'. This session emphasized the importance of choosing your priorities wisely. There was also a session on sexuality, presented by Catherine Mathieu, who illustrated that pleasure and intimacy are still accessible despite illness. A last session entitled 'Empty your heart' was held for groups of patients and caregivers separately, to allow individuals to share their feelings with others in their position. The first conference day ended on a high note with comedian Johanne Fontaine, who demonstrated that we're able to laugh and appreciate life even when it's difficult.

On Sunday morning, we had the honour of hosting Marie-Sol St-Onge and her partner Alin Robert. After a horrific encounter with flesh eating disease, Marie-Sol had to undergo amputation of both of her arms and legs. Marie-Sol and Alin's love and their enormous courage through this time of trial powerfully demonstrated that we CAN 'choose happiness, despite life's challenges'.

The conference ended with the general assembly of the Association. After the elections took place, we were pleased to announce our new Board members:

Alain Chabot from Lévis
Hugues Boulanger from Plessisville
Dolorès Carrier from Plessisville
Michel Anctil from Plessisville
Jean-Pierre Vigneault from St-Romuald
Judith Moatti from Montrèal
Denis Cormier from Plessisville
Pierre Lachance from St-Hyacynthe
Jacques Gariépy from Victoriaville

Our Board is united in the PHight against PH, and we look forward to a fruitful 2015 year.

Contributed by: Dolorès Carrier, HTAPQ Board Member

Connections submissions

The deadline for submissions for the next issue of Connections is July 15th, 2015. **Connections is your publication.** Tell us about your support group, recent event, share your story or tell us about a phenomenal caregiver in your life. Let us know how you cope with PH: how you live and work every day. We'll accept articles, personal PH stories, quotes, photos, tributes, poems, drawings and more for consideration in the magazine. If you're not comfortable writing your story, contact us, we'll interview you and write the story for you. This is Canada's PH community's newsletter: let your voice be heard!

Please send submissions including your contact information (phone and/or email) to:

Newsletter submission
PHA Canada
Suite 208, 1311 Howe Street
Vancouver BC V6Z 2P3

Or email it to "Newsletter Submission" at info@phacanada.ca!

Work submitted will be printed as space permits.

Connections e-Magazine!

Did you know that Connections Magazine is also available in an electronic format? If you would prefer to read a PDF version of Connections instead of the printed version, we would be pleased to send it to you electronically. Please email us at connections@phacanada.ca to let us know.

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.