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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Special Issue: PH & Family Planning

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

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In this Issue:

Inside PHA Canada

Your Community

Family Planning Feature

Research Corner

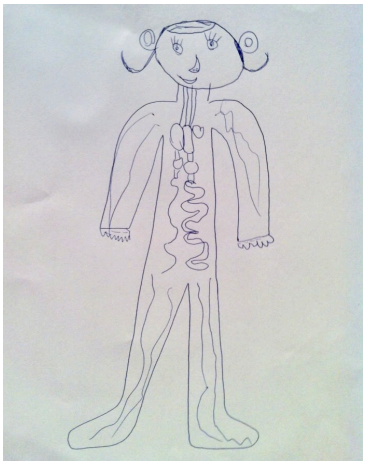
Message from the Chair:

Understanding Current and Future Parents Affected by PH

It's hard to believe that I'm already writing for our Winter issue. We've come such a long way since last Winter. Is it just me or is time flying by, yet going slowly at the same time? With 2020 behind us, my hope is that you are all continuing to stay safe and that there are brighter days ahead for this New Year. Give yourself a pat on the back for getting through a very challenging time.

This issue of *Connections* has a special focus on family planning. Whether you're a PH patient, a caregiver, or a family member, we all know family planning can be a touchy subject. PH patients can experience difficulties in various ways, whether they want to have a baby or already have children upon diagnosis. PH can derail all plans when it comes to family life. PH patients and their families go through emotional turmoil, which is an added stress to having the disease. I've met childless women—some by choice and some unwillingly because of PH—who struggle with having to answer to people who ask: "are you planning on having kids?" or "when are you having kids?". Some people just don't realize how painful such questions can be.

While I already had kids when I was diagnosed with PH, my daughters were only three and five years old. Life suddenly presented many challenges. Some people choose to keep some aspects of their illness private, but I always felt it was important to be honest with my kids. They knew right from the beginning that their mom was sick. I remember drawing a picture to show my girls what was going on with my lungs and heart. My oldest daughter then drew a picture of the human body to help her understand further.



Carys, age 5 (2013), draws the human body so she can understand what's going on with my lungs. We had a lengthy chat.

My husband, Darren, and I also felt it was important to continue on with life as if it were "normal".

> **"As much as a PH diagnosis presents new challenges, we still wanted to continue doing the things we loved; namely, travelling."**

And so, that's just what we did. I no longer work as an elementary school teacher, so we've taken every opportunity to travel and explore the world. I really wanted my girls to have a bunch of travel memories that included me.

In this issue you'll also be introduced to PHA Canada's new strategic plan (page 8). Thank you to everyone who provided input and helped us refine our vision for the next three years. Part of this plan includes better understanding the lives of those affected by PH in Canada and so, in early 2021, we'll be launching our *Canadian PH Community Survey* (page 31). The survey results will allow us to better understand the Canadian PH community and will also give us an opportunity to focus on areas that most need improvement. We hope you will participate in the survey. Your feedback is always so important to us.

Be well and stay safe,

Nicole Dempsey, Living with PAH since 2013
Board Chair, PHA Canada

Message from the Executive Director:

Choosing Generosity in 2021

I love celebrating the New Year – it's one of my favourite times of year. I enjoy the reflectiveness that December inspires and the quiet darkness of January. I try to embrace the spirit of hope and possibility that a new year brings. Of course, this year brings even more anticipation for better days ahead than most. Nonetheless, if 2020 has left you feeling weary or sorrowful, I understand and hope you find some comfort in knowing you are not alone. I hope this Winter issue of *Connections* provides a warm reminder of the support that exists for you every day of the year (page 6) and the very special community you belong to (page 10).

Being part of a community of peers who can relate to our most intimate struggles is immeasurably valuable. Few things in life are more intimate than the decisions we make (or don't make) and the actions we take (or don't take) when it comes to having a family. "Family planning" can be deeply personal and pretty complicated at the best of times. So, when questions of family planning intersect with our health status, "complicated" is often just the tip of the iceberg. Decisions you might have reasonably expected to be yours alone or made with a partner, suddenly become dependent on the judgments and actions of others.

> **"What you thought would be a mostly private affair, can instead involve teams of experts, intricate legal arrangements, and a profound dependence on the generosity of others."**

This special feature on *PH & Family Planning* is very personal for me, because I know what it's like to be told that I couldn't get pregnant because of a chronic health condition. (Not a life-threatening condition, but one that caused me chronic pain for 20 years and went undiagnosed until it made me infertile.) I know the difficult conversations that come with weighing your options. And then the even more difficult conversations about what happens if you run out of options. I know the immense privilege of even having options, including medical options that didn't exist when I was conceived. I know the disappointment of learning that infant adoption was never going to happen for us and the complexity of bringing a child into this world in an unconventional manner. I also know the overwhelming pride and gratitude that comes with bringing a baby you weren't "supposed" to have into this world.

As you might imagine (especially those of you who know me), I was unable to read the stories and quotes shared in this issue without a lot of tears (pages 18-23). Like me, many of you will relate to the heartache and determination that is part of the journey to parenthood. Or perhaps you will find solace in the reminder that parenthood is not the only path to family and finding our place in this world.

> **"Either way, since few things in life are more intimate than the making of our families, I have immense gratitude for the people who shared their stories with us for this issue. It is a special act of kindness to make one's private life public for the benefit of others. Thank you for being so generous and open with us."**

The last couple of years have taught me a lot of about what it means to be generous and my hope for 2021 is that we will all find more ways to be generous. For it takes an abundance of generosity to make good things happen in this world! It's remarkable to think about the amount of time and effort that has gone into a small community like ours raising $100,000 for research in only five years (page 30). It's awesome to think about the hard work that goes into developing a possible new drug therapy (page 28) or the thoughtfulness that goes into understanding the patient perspective in PH (page 29). I know the PH community has a lot to give and believe in 2021 you will give more generously than ever. I hope you will start by accepting our invitation to fill out *PHA Canada's Canadian PH Community Survey* (page 31) and continue by joining with us virtually throughout the year to learn, advocate, raise awareness, and support one another through these extraordinary times.

With Gratitude,

Jamie Myrah
Executive Director, PHA Canada

MEMO: INSIDE PHA CANADA AT A GLANCE

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

Going Virtual

The global COVID-19 pandemic has emphasized the significance of maintaining community connection, even if it has to be virtual. Although we had to postpone our plans for a community conference in 2020, PHA Canada made sure to create occasions for the PH community to connect with experts and with one another. Since August, PHA Canada has hosted webinars with community leaders to discuss questions and concerns about four important topics: the return to school for children with PH, managing mental health in the context of PH, making nutrition decisions when living with PH; and understanding PH treatment options. View all PHA Canada webinars at phacanada.ca/Webinars.

COVID-19 & Back-to-School: A Q&A for Families of Children with PH

In August, PHA Canada held a webinar about the return to school for children with PH, in the context of COVID-19. A panel of pediatric PH medical experts answered questions from PH parents and other caregivers. The panel focused on giving parents general information and tools to make decisions according to their own personal circumstances.

The panel featured:

- Janette T. Reyes, Nurse Practitioner, The Hospital for Sick Children Cardiac Clinic, Toronto, ON
- Dr. Andreea Dragulescu, The Hospital for Sick Children Cardiac Clinic, Toronto, ON
- Susan Richards, Nurse Practitioner, Stollery Children's Hospital, Edmonton, AB
- Dr. Anne Fournier, CHU Ste-Justine, Cardiology Department, Montréal, QC

Janette T. Reyes, Nurse Practitioner, during the *COVID-19 & Back-to-School* webinar

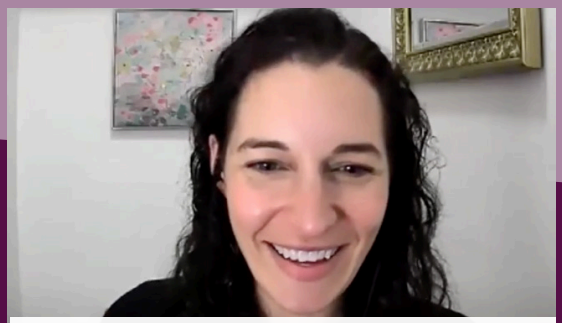
Fall Education Series

PHA Canada was pleased to present an online education series in the Fall. The first webinar in the series—*Facing Mental Health Issues*—was a follow up to the Summer 2020 issue of *Connections* magazine. The session featured fictional stories of PH families that explored some of the reasons why people affected by PH are at an increased risk of facing mental health challenges such as depression and anxiety. A panel of mental health experts—including PAH patient and retired counsellor Jeannette MacKeen—shared their thoughts and advice on how people can approach life's challenges in a healthy way and build resiliency.

The second webinar featured Registered Dietician Sylvia Rinaldi discussing her important research: *Nutritional Status of Patients with PH*. Sylvia is also a clinical researcher and recipient of the 2016 and 2017 *PHA Canada Paroian Family PH Research Scholarship*. Check out her webinar to learn more about the nutritional concerns of people living with PH and how they can improve their diet.

The third webinar—*Treatment 101: Introduction to PH Treatment*—was about understanding PH treatment options so that PH patients can be active participants in their own care. PHA Canada Executive Director Jamie Myrah, as well as Lyda Lesenko and Jessica Pinto, PH Nurse Coordinators from Montréal, QC, spoke about treatment options for PAH and CTEPH, side effects and the importance of medication adherence, and the future of PH therapies in Canada.

Watch the entire webinar series here: phacanada.ca/Webinars.



Joanne Schwartz, Social Worker and Counsellor, Vancouver PH clinic, BC, *Facing Mental Health Issues*, October 2, 2020

PH Buddy Program

Make new connections over the phone or virtually by chatting with someone from the PH community who understands what you're going through. Sign up to become a "PH Buddy". Buddies will help one another stay safe and socially connected during this uncertain time.

Visit phacanada.ca/PHBuddy.

PH Blog

Read the latest blog posts from healthcare professionals, researchers, advocates, and people living with PH. Read about diverse subjects such as the quality of life of PH caregivers, how pets can help on a PH journey, the importance of the "patient perspective", and updates on PHA Canada's advocacy efforts, PH research, COVID-19, and more.

Visit phacanada.ca/PHBlog.

COVID-19 & PH

Visit our dedicated webpage for trustworthy information about COVID-19, as well as education and support resources, including a special bilingual Q&A session with Dr. Sanjay Mehta, MDCM, FRCPC, PHA Canada Founding Board Member, Eternal PHriend, & Past-Chair, and Director of the Southwest Ontario PH Clinic, London, ON.

Visit phacanada.ca/COVID-19.

Online Store

It's now easier than ever to purchase your favorite PHA Canada item from our brand new online store! Sometimes spreading PH awareness is as simple as slipping into a comfortable t-shirt or hoodie, wearing a beautiful mask, or even putting on cozy mittens. Your purchase shows support for the PH community, while also providing a financial contribution to PHA Canada.

Shop here: phacanada.ca/Store.

For Family Planning resources, go to page 26.

PHA Canada's Annual PH Community Conference is Going National!

This year, PHA Canada brings you a COVID-friendly conference experience from the comfort of your own home. Join us in June for our first **bilingual and virtual** *National PH Community Conference*!

We are also pleased to host the *2nd National PH Medical Think Tank* for PH medical specialists. For the safety of all, this event will also take place remotely this year.

Save the Date:
***National PH Community Conference* begins June 12, 2021**
***2nd National PH Medical Think Tank* returns June 10-11, 2021**

Stay tuned for more details of a month full of learning, connection, and inspiration.

Visit phacanada.ca/Events for all the updates.

2021–2024 PHA Canada Strategic Plan

The Pulmonary Hypertension Association of Canada's strategic plan reflects the expanding cohesiveness of our national community, our developing sophistication as an organization, and our enduring commitment to improving the lives of Canadians affected by PH.

GOAL #1: All patients and caregivers in Canada live well with pulmonary hypertension.

- Objective 1.1: Empower PH patients and caregivers to be active participants in their care.
- Objective 1.2: Facilitate connections within the PH community that foster peer support and mentorship.
- Objective 1.3: Improve access to resources and supports for underserved members of the PH community.

GOAL #2: PH patients have equitable access to timely and optimal care across Canada.

- Objective 2.1: Enhance understanding of the diagnosis and management of PH among Primary Care Physicians and other health care providers.
- Objective 2.2: Canadian PH patients have timely, consistent, and universal access to all Health Canada-approved treatment options.
- Objective 2.3: Facilitate the implementation of standards of excellence for PH care in Canada.

GOAL #3: Canadian PH research advances the care of patients and brings us closer to a cure.

- Objective 3.1: Grow our investment in Canadian PH research.
- Objective 3.2: Promote awareness of the excellence of Canadian PH research among the PH community and general public.
- Objective 3.3: Build the capacity of Canadian PH patients to participate in research.

GOAL #4: PHA Canada is recognized as the national leader and voice of the Canadian PH community.

- Objective 4.1: PHA Canada is a trusted source of information for the Canadian PH community.
- Objective 4.2: PHA Canada is a trusted source of information about the Canadian PH community.
- Objective 4.3: PHA Canada is a trusted partner of key stakeholders in PH patient care, education, research, and advocacy.

GOAL #5: PHA Canada has sustainable financial and human resources to meet the future needs of the Canadian PH community.

- Objective 5.1: Grow individual and corporate support while diversifying revenue streams.
- Objective 5.2: Staff and Board have the capacity to make continuous organizational improvements.
- Objective 5.3: Optimize technological solutions to create efficiencies and improve impact.

Your Community

This year, community events could not take place in person because of the measures and restrictions in place to prevent the spread of COVID-19. Instead, PHA Canada and the PH community rolled up their sleeves, learned new technologies, and organized virtual events!

Thank you to the community leaders and volunteers who support the PH community in difficult times and to the fundraisers and donors who create a better life for all Canadians affected by PH!

Your Community In Action

November PH Awareness Month 2020

November PH Awareness Month 2020 was all about bringing positivity and connection, even if we couldn't be together physically. The Canadian PH community exceeded all our expectations with their involvement and support. Community leaders shared their stories with us on Facebook Live, volunteer hosts supported the PH community during virtual meet-ups, and fundraisers and donors helped us raise more than $35,000!

Facebook Live

In Conversation with...

PHA Canada Board Chair Nicole Dempsey and Executive Director Jamie Myrah went live twice: first on November 2nd to kick off PH Awareness Month and then again on November 30th to wrap it up;

Jane Sernoskie, living with PAH since 2016, answered questions about her journey with PH, the important connections she's created with fellow community members, and how PHA Canada has helped her along the way;

Natalie Roy answered questions about the double-lung transplant she received in 2017 after living with PAH for 12 years, and an important relationship that helped her get through it all;

Wendy Bedard, PH Caregiver since 2010, shared her journey through caregiving for her husband Kendall, living with PAH, and how her friendships with members of the PH community has been an important source of support;

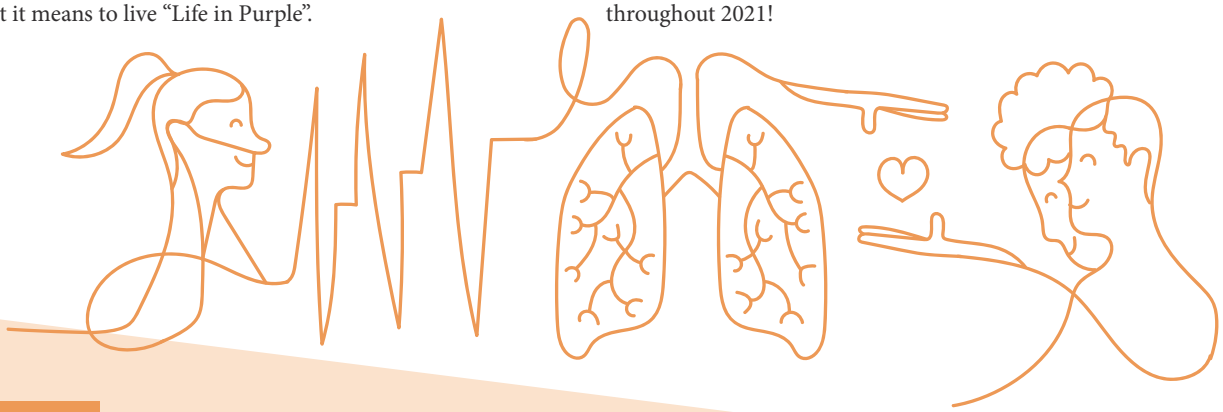
PHA Canada Board Co-Vice-Chair and Director of the PH Program at the Ottawa Heart Institute Dr. Lisa Mielniczuk talked about advocating for PH patients and how important it is to connect with her patients and their caregivers.

Watch all of November's conversations here:
facebook.com/PHACanada/live_videos

Virtual Meet-Ups

PH patients, caregivers, and health care professionals each had a chance to gather online with their peers to celebrate PH Awareness Month. These casual meet-ups were a chance to connect with PHriends new and old, the ones that really know what it means to live "Life in Purple".

Using the magic of Zoom-technology, participants were able to join small groups led by volunteer hosts from across the country, all from the comfort of home. We received lots of great feedback about the meet-ups, so stay tuned for the chance to do it again throughout 2021!



Celebrating Our Major Fundraisers

Virtual 8th Annual Ottawa *6-Minute Walk for Breath*

Since 2012, PH Nurse Carolyn Doyle-Cox, the Ottawa Pulmonary Hypertension Support Group, and PHA Canada have put on a fundraiser called the *6-Minute Walk for Breath*. Every year the Ottawa PH community gathers to raise funds and awareness of PH. This special event is designed to educate participants on what it's like to live with PH using the famous "six-minute walk test". This year, when it was clear the community would not be able to gather in Ottawa, the event became a virtual walk and we invited people to join in from anywhere in the country. Participants from coast to coast walked for six minutes, many while wearing a mask and holding weights in order to get a feel for what it's like for PH patients to do the walk.

On Saturday, November 7, 2020 organizers from Ottawa, Ontario welcomed PH supporters from across Canada to the Virtual 8th Annual Ottawa *6-Minute Walk for Breath*. Together participants raised an incredible $20,000!

PHA Canada Board Director Emily Pinckard, living with CTEPH since 2017, was one of our top fundraisers. Pictured with her partner Josh Perlstein and her service dog Oscar in Oakville, ON.

Denise Rumbolt walked with her family in memory of her daughter Candice Cooper in Happy Valley-Goose Bay, NL. Candice passed away in 2015, at age 21, after being diagnosed with idiopathic PAH in 2014.

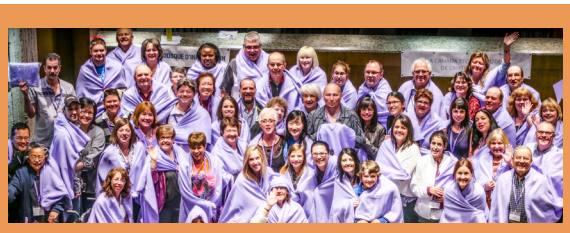
Sadie walking with her mom Jamie Myrah, PHA Canada's Executive Director, in Vancouver, BC.

Brinley Marks, diagnosed with PAH in 2020, walked with her best friends, Brandee Robertson-Grafton and Augustus, Brinley's service dog, in Sherwood Park, AB.

Corporate Donors

A special thank you to McKesson Specialty Pharmacy for once again making a $3,000 matching donation in honour of November PH Awareness Month.

Chicago Title Insurance Company Canada matched all donations made in November to their annual employee giving campaign.



Create your own personal fundraiser any time of year at **phacanada.ca/Fundraise**

Major Donor Recognition

The most important donors an organization can have are the ones that stick with you year after year. In this special issue we pay tribute to our most dedicated donors who – over the past five years of giving – have had a major impact on the lives of Canadians affected by PH. It is because of your generosity that no one in Canada has to face a diagnosis of pulmonary hypertension alone. Thank you for placing your trust in PHA Canada and for your commitment to creating a better life for all Canadians affected by PH.

Individual Donors

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PHA Canada's Most Frequent Donor

As of January 2021,
we celebrate
Jas James's
50th donation!

Follow her lead and
become a monthly donor:
phacanada.ca/Donate

Fondation HTAPQ News

2020 and the beginning of 2021 will be remembered all across the globe. Closer to home, in Québec, people with pulmonary arterial hypertension (PAH) and their loved ones have experienced nearly a year of lockdown and worry. All public health measures have brought their share of questions, insecurities, and anxiety. Dolorès Carrier, President of the pulmonary arterial hypertension foundation of Québec (Fondation de l'hypertension artérielle pulmonaire du Québec) updates us on their activities.

New Opportunities

In order to reassure PAH patients, the Fondation took the opportunity to organize conferences on Zoom, a digital platform. As a result, members of the Fondation had access to several specialists. A doctor, nurse, psychologist, therapist, and psychotherapist took turns informing and reassuring participants. We even held our Christmas party on Zoom! Even with social distancing, we all enjoyed these happy moments and even had a visit from Santa Claus!

This has given us a chance to reach people regardless of distance. Everyone had the opportunity to participate safely in conferences from the comfort of their homes. Before each conference, there was a greeting period during which people could chat and thus feel less isolated.

That being said, we must remember that some of our members do not have access to the internet. We have to find ways to reach them, especially since they are the most isolated.

Awareness

The Fondation collaborated with PHA Canada to raise awareness of PAH during PH Awareness Month in November. In Québec, 20 patients agreed to make a video clip explaining the symptoms and the consequences of the disease, what it's like to live with it, etc. This campaign was an overwhelming success. The clips were posted on Facebook and viewed by thousands of people. Unfortunately, early diagnosis of the disease is still a major issue. Still today, patients are transferred from one doctor to another before receiving the right diagnosis. The disease has time to progress and patients often see their quality of life deteriorate in an irreversible manner. Awareness campaigns are therefore necessary among the population and the medical profession.

Fundraising Campaign

Like most organizations, the Fondation experienced a significant decrease in funding due to the pandemic. Not wanting to put the health of our volunteers at risk and wanting to respect public health rules, we cancelled our fundraising campaigns. However, no-contact campaigns were successfully launched. Surprisingly, we also received more donations than usual. Hopefully, we will be able to resume our fundraising campaigns so that we can continue to provide direct financial assistance to those in need.

Board of Directors

The members of the Board of Directors had no choice but to hold their meetings on Zoom. Even if this new way of doing things required some adaptation, it is here to stay. It saves us a lot of time by avoiding travel and it allows people living in remote areas to readily participate in meetings. The general meeting was even held by videoconference. However, this is a formula that will never replace face-to-face meetings, which we will resume as soon as it becomes possible.

The Board of Directors is composed of nine members. I have the good fortune to be working with caring people. Together, we want to improve the quality of life of people with PAH, in Québec.

Contributed by: Dolorès Carrier, President, Fondation HTAPQ

First row: Dolorès Carrier, President; Pierre Lachance, Vice-President; Renée Levaque, Secretary

Second row: Pierre Gagnon, Treasurer; Stéphanie Théorêt, Board Member; Jean-Pierre Vigneault, Board Member

Third row: Line Ducharme, Board Member; Denis Cormier, Board Member and Founding President; and Judith Ross, Board Member

Special Feature: PH & Family Planning

A PH diagnosis can mean accepting a lot of new limits, including limitations to one's choices for how and when to build a family. Our special feature explores both the medical realities of PH and pregnancy/birth, as well as the lived realities of four courageous women whose family plans got turned upside down by PH.

> **"Family and friends, who I would see around, would never have known the internal turmoil I was dealing with, facing a future devoid of motherhood."**

PAH and Pregnancy

Pulmonary arterial hypertension (PAH) is a complex disease with significant health implications. Due to the nature of the disease, it is recommended that pregnancy be avoided in this patient population. Recognizing the harmful effects during pregnancy is of critical importance to understanding the reasons for this recommendation.

Women of child-bearing age make up a significant proportion of the PAH population. The decision to plan a family is relevant among this group of women, and in the era of access to good PAH-targeted therapy, women are living better and longer so therefore might consider planning a pregnancy as a feasible option. Certainly, there are examples of successful pregnancies among the Canadian PAH community.

The Risks of Pregnancy in Women with PAH

Historically, before the availability of PAH-targeted therapy, pregnancy was associated with extremely high risks to both mother and fetus; with a third to half of women dying during or shortly after pregnancy. While outcomes have improved in the modern era of treatment and pregnancy care, the risk of critical outcomes such as death or urgent lung transplant remain unacceptably high at 11%–33%.

Patients are not all equal in their degree of risk. Women with severe PAH when they become pregnant have worse outcomes than those who have milder and more stable disease. But even in the most well controlled patients, disease can worsen over the course of pregnancy. The greatest risk to mother is early following delivery, where most deaths occur. There is also increased risk of poor outcomes in baby, with a death rate of up to 13%. Most fetal deaths are because of maternal death prior to delivery, compared to live-born infants who are also at risk of premature birth and growth retardation.

> **Because of the unacceptably high risks to mother and baby, current PAH guidelines strongly recommend that pregnancy be avoided in patients with PAH, and that women who become pregnant be offered the option of having a therapeutic abortion.**

The Changes in the Circulation due to Pregnancy

There are many changes that occur to a woman's body to support the pregnancy. Within the cardiovascular system, the heart and circulation must support both the mother and the growing fetus. To accomplish this, the volume of plasma (the fluid part of the blood) increases by one half through to the end of the third trimester, but changes start soon after the pregnancy is established. The amount of red blood cells also increases over the course of the pregnancy.

With the increased blood and plasma, the heart must also increase its ability to circulate the extra volume. This is accomplished by increasing the heart rate. Together, the increased heart rate and blood volume would increase the pulmonary arterial pressure; however, in normal pregnancy, there is compensation to adjust for the high flow state. During pregnancy, the amount of blood being pumped by the heart increases. This increase is due to hormonal effects as well as an increase in the amount of blood in circulation. In healthy individuals, the lung's circulation can adapt to this change. However, women with PAH cannot adapt to this new demand on the heart. This will lead to additional stress on the right heart and potentially reduce the ability of the heart to meet the needs of the mother and baby for normal development.

This causes an increase in pulmonary arterial pressures, which places further stress on an already strained right heart. This in turn leads to worse symptoms (shortness of breath, fainting, fatigue), signs of a failing heart (leg swelling), and ultimately risk of death from heart failure.

Labour and delivery introduce further stress on the system because of the respiratory efforts and straining during contractions. This is compounded by swings in pressures and return of blood from the uterus into the maternal circulation. Following delivery, there is a rapid drop in hormone levels and volume status, increased resorption of fluid, and risk of blood clots, all of which may contribute to a sudden worsening with progression in disease and the highest risk of dying.

Recommended Management

Recognizing the increased risks to women with PAH during pregnancy, sadly, avoidance of pregnancy is still recommended. For those who have become pregnant, the guidelines recommend termination of the pregnancy as the safest strategy. This should ideally be performed in the first trimester at an experienced center.

For women who choose to continue, the pregnancy is considered "high risk". Management should be provided in an expert PH center with multidisciplinary involvement in the delivery of care. This can include a PH specialist, a PH nurse, a cardiologist, an obstetrician, a maternal-fetal medicine specialist, an anaesthesiologist, and a neonatologist. Care is focused on both the mother and the growing fetus.

> **For those who are still considering pregnancy, they should discuss the risks with their PH team.**

Women who are pregnant and wish to continue with the pregnancy can expect frequent clinical evaluations and testing to ensure the well-being of both mother and baby. Within the PH clinics, testing of standard parameters such as clinical and physical examination, six-minute walk testing, lab testing for BNP, and echocardiography continue. The frequency of visits increases as pregnancy progresses. As a general guide, recommendations are that women be evaluated monthly in the first trimester, every two weeks in the second trimester, and weekly in the third trimester for an assessment of the status and stability of their PH.

Women can also expect to have visits with the obstetrician, as well as regular examination of the baby with serial fetal ultrasound assessments to assess for fetal growth and development.

In stable women, delivery is usually planned around 34–36 weeks of gestation at an expert center. Elective caesarean section is preferred, as vaginal delivery is associated with harmful cardiovascular demands of labour. Women are monitored closely in an intensive care setting throughout delivery and post-partum for at least several days.

PAH Medications

Most women with PAH require medications to manage the disease prior to pregnancy. Not all medication is safe to a developing baby and must be adjusted when pregnancy is determined. Medications considered dangerous to the developing baby include warfarin for prevention of blood clots and many PAH-targeted medications: endothelin receptor antagonists (ambrisentan, bosentan, macitentan) and soluble guanylate cyclase inhibitor (riociguat). Risks of oral prostacyclin IP inhibitor (selexipag) are unknown, but avoidance is suggested.

Medications considered safer include calcium channel blockers (amlodipine, diltiazem, nifedipine), phosphodiesterase 5 inhibitors (sildenafil, tadalafil), and prostacyclin analogs (epoprostenol, treprostinil). A change in treatments to safer alternatives poses an inherent risk of disease destabilization during the change. Many patients will require an increase in therapy. IV epoprostenol is frequently used as it enables more accurate titration of dosing as needs change when pregnancy progresses, giving significant advantage during labour and following delivery.

Other Considerations

There are considerations beyond the issues identified above. Many women live a far distance from the PH centers that have the knowledge and expertise to manage high-risk patients. Therefore, if there is a sudden change, local hospitals may not be able to safely care for the complications that could arise. Additionally, guidelines may suggest very close monitoring and testing of patients, but the reality of limited resources in our health care system means that this may not be easily available.

Finally, having a baby is more than the direct medical care through the pregnancy. It involves support from all family members who will be involved in raising the child. One must consider the possibility that the mother may have worsened PAH or death following the pregnancy. The family must be accepting of this possibility, both in the care of the mother and the child. PAH is also a hereditary disease for individuals carrying a predisposing genetic mutation. Certainly, gene mutations have been identified to date to inform of this risk in advance.

The decision to plan a family or continue with an unplanned pregnancy in women with PAH has challenges that are very difficult. There are many facets of the decision that must be carefully considered and individualized. Unfortunately, in 2020, PAH remains an incurable progressive severe disease with risks considered unacceptably high in pregnancy, leading to the guideline recommendation for avoidance.

Contributed by: Drs. Kristina Kemp and John Granton

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CTEPH and Pregnancy

Chronic thromboembolic pulmonary hypertension (CTEPH) is another cause of PH that may be diagnosed in women of childbearing age. In contrast to PAH, some forms of CTEPH may be successfully treated with interventions such as pulmonary endarterectomy surgery (PEA), balloon pulmonary angioplasty (BPA), or other medical therapies. Pregnancy in women with CTEPH has some unique considerations.

Pregnancy outcomes in the CTEPH population are not well described in medical literature. All patients with CTEPH require life long use of a blood thinner. Warfarin and oral anticoagulants (apixaban, dabigatran, edoxaban, rivaroxaban) are not recommended in pregnancy; a switch to a safer alternative such as heparin is required. CTEPH patients who are considered "cured" post-PEA/BPA no longer have PH, therefore have reduced risk. Nonetheless, the pregnancy is still considered "high risk" and would benefit from close monitoring by a multidisciplinary team. There are reports of successful pregnancies in this population.

In contrast, women with CTEPH who are not eligible for curative interventions, or in those who have evidence of persisting CTEPH despite interventions, are typically managed medically with riociguat. Unfortunately, riociguat is known to cause harm in the fetus and pregnancy is therefore contraindicated. There are no other approved alternative CTEPH medications presently available in Canada. For newly diagnosed CTEPH patients, where pregnancy is already established, PEA and BPA interventions are very high risk to the fetus and would often be delayed until the pregnancy is completed. This delay however also places mother and baby at high risk.

Planning a Pregnancy While Living with Pulmonary Hypertension

Lea George was diagnosed with PAH in 2016, when she was 32. In 2018, after consulting with multiple specialists, Lea and her husband Ryan decided to make a detailed pregnancy plan they believed would lead to a safe outcome. With the help of a diverse medical team, they got their wish when their son Ethan was born in April 2020.

Note: If you have PH and are considering becoming pregnant, it is strongly recommended you consult with your PH team as soon as possible.

Lea and her son Ethan

After the shock of being diagnosed with pulmonary arterial hypertension (PAH) wore off, the hardest part of living with the disease was being told I could never become pregnant. My fiancé, Ryan, and I were actively trying for a baby and ready to start our family.

I felt alone in my difficult and unique situation. We put ourselves on the adoption list and were told it could be eight years long. We also investigated IVF and surrogacy options, and we found both could be too risky.

The longer I lived with PH, the more comfortable I felt. The data said there was a possibility of death to a PH patient, but I felt I was well enough to carry a pregnancy and be able to take care of the baby afterwards.

I approached my PH specialist, Dr. Krista Kemp, and not surprisingly, she presented me with the many risks. She was successful in talking me out of it the next few times I got the courage to broach the subject. Through a PH Facebook group I heard out about Dr. John Granton, a PH specialist in Toronto. He had seen pregnant women with heart complications, and I thought he would be an ideal person to talk to. I asked Dr. Kemp for a referral to see him. Due to logistics, we talked to Dr. Granton via teleconference (us at the Moncton Hospital and him in Toronto). He was very relaxed and open-minded about discussing the issue and seemed quite amazed at how well I was doing. Ryan was finally onboard with the idea. Dr. Granton sent me for some more tests and, looking at my results, thought I was a good candidate to carry a baby.

Dr. Kemp took over and we started our detailed pregnancy plan in October 2018. I switched from Opsumit™ to Adcirca™ and had tests to ensure my heart functions and pressures were stable. We were given the green light to start trying in June 2019. In August I took a pregnancy test, and it was positive!

My weeks were filled with appointments; baby and I were followed closely by a whole team of specialists. I was seen at the high-risk maternal fetal clinic. One positive that came out of being a high-risk pregnancy was that I got to have so many ultrasounds. Luckily, my pregnancy was uncomplicated; I was even able to walk 640 meters on my six-minute walk test at eight months pregnant! I worked full-time at the Moncton Hospital until the end. I loved every second of my pregnancy—the nausea, heartburn, and all. I just felt grateful to experience it.

Throughout this whole journey my PH nurse, Kelly Gould, was there. She attended most of my appointments to bridge the PH side with the obstetrics side. She was so supportive.

> **"PH and pregnancy are a taboo subject in the PH world. I felt like a horrible person for constantly bringing up the idea, like I was doing something wrong. When I was pregnant, I kept it all very quiet because I feared judgement."**

I knew what I was doing could be viewed as irresponsible. Kelly always reminded me that I was the only one who could live my life.

It was decided that a c-section would be the safest method of delivery, so it could be in a controlled manner. They planned for me to deliver at 37 weeks when the baby would be developmentally complete. I would go to Saint John, New Brunswick to deliver at the heart center, which would be better equipped to handle my case.

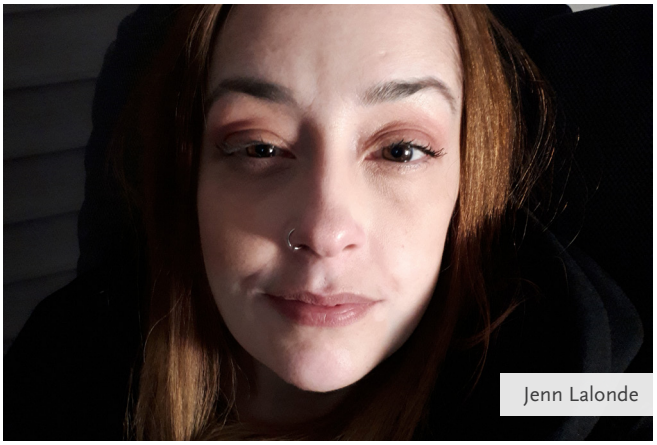
Our baby was scheduled to be born April 3, 2020. We drove to Saint John and were admitted the evening prior. We were waiting to find out the baby's sex, so it was all very exciting. We were so relaxed during the entire pregnancy, which we didn't think could be possible with my condition. We had total confidence in our team. Ethan Ryan Blackbeard was born at 9:25 am the following morning weighing 7lbs and 1oz. I had no PH related complications and the COVID pandemic was really the only stress on anyone's mind. It was nothing at all how I pictured our delivery so many times in my head.

Ethan had to go to the Neonatal ICU after he was born (where he spent a week) and I had to go to the CCU to be monitored. So after delivery, Ethan went his way and I went mine. When they put him on my chest for the first time 36 hours later it was the most perfect moment. I could feel us both just take a deep breath and settle. All the pain disappeared in that moment. We were so content and have been so ever since. He is such a wonderful baby; we are so fortunate. We cannot imagine our life without him. He has brought us and our families so much joy. I plan to watch him discover and grow for a very long time thanks to the two greatest PH doctors—Dr. Kemp and Dr. Granton, my PH nurse Kelly, and the rest of our amazing teams at both the Moncton and Saint John hospitals.

Contributed by: Lea George, Living with PAH since 2016

Making the Journey from Grief to Peace

Jenn Lalonde was diagnosed with PAH in 2003, when she was in her mid-twenties. She writes about her journey to acceptance after years of remorse over not having children. In sharing her story, she hopes to show those newly diagnosed that finding peace is possible.



Jenn Lalonde

I was diagnosed with pulmonary hypertension in May of 2003. At the time, I was 24 and in my first serious long–term relationship. As far as I can recall, we had not discussed the possibility of having children that early into our relationship.

Before my diagnosis, we were both young and in love. Things were headed in the right direction in our lives: we both worked full time; we had just bought our first house; we were living and having fun. Things were looking up. As May of that year rolled around, I had been through a ringer of hospital tests to see why I couldn't climb a flight of stairs without having to sit and catch my breath. I was eventually diagnosed and life as I knew it changed before me.

I was quickly told that I wouldn't be able to bear children. I would likely not survive the childbirth and my disease could worsen if I were to get pregnant and carry a baby.

This news was quite shocking, even though I had never been one to dream about having children or to picture myself as a mother. A close friend offered to be our surrogate, but we would have had to act soon as she was nearing 40 and didn't want to have another baby that late in her life.

I was always grateful for that offer, but I couldn't make that kind of a decision at such a stressful time. I was not ready. I was dealing with the shock of a life-changing diagnosis, extreme depression, and a complete change of life. It would not have been the right time for anything, let alone a baby. I had to heal a bit and come to terms with my diagnosis. I know that many PH patients in my situation have considered adoption, but it wasn't ideal for my situation. I did, however, take on a puppy.

For a few years, it was very difficult answering to people about why I wasn't getting pregnant. That was 17 years ago, and some people did not have the decency to respect boundaries. Many would ask rude, invasive, and senseless questions. It was very difficult, being that young and facing a potentially terminal illness. I've always been rather bold, so eventually I reached a point of no longer caring what anyone thought and I would answer simply and truthfully: "I can't carry a baby because I might die."

Everything took its toll. PH sunk me into the deepest depression.

> **Dealing with the idea of never having children was burdensome, despite the fact that I didn't want children. The fact that I could not have them tore me apart somehow. I grieved for that child I could not have.**

Every time someone close to me got pregnant, I cried before I could feel excited about it. I mourned the fact that I couldn't have that life. I was consumed with jealously around this topic. It felt like a constant stab in the back whenever it was talked about or brought up. I was not in a great place and there was some time, in my early 30s, when this was all I thought about.

I am now 41 and I have healed a lot. I no longer get grief stricken if a friend gets pregnant. The friends I do have (most with kids), I am very close with. I love spending time with them. Since I'm a big kid at heart, we have tons of fun when we are together. I now feel completely at ease and at peace with the fact that I will never be a mother. Still, I do have guilt for never being able to give my parents the opportunity to be grandparents, even though they have never once mentioned it. I think they are fine with it too.

I have witnessed and helped with the births of all four of my cousins' babies. I even cut each umbilical cord. I can hardly express how much this has meant to me and helped me. This certainly isn't something I will ever get to be a part of with anyone else, so it has significant meaning to me. I will be forever grateful for these experiences.

I sometimes imagine what my life would have been like with a child: What would my child have been like? Looked like? I think about "this child" at different ages and in different circumstances. I can do this, now, without the gut-wrenching resentment I once lived with.

While I do not currently have a dog, I am content with someday adopting. Animals bring me a great sense of joy and I look forward to someday finding a dog who is the right fit.

> **My hope for all those young, newly diagnosed PH patients—given the hard news about bearing children—is that they can find some peace with it. There are always different options out there if they choose. It might take a few years to obtain that peace, but they should know that it's certainly attainable.**

Contributed by: Jenn Lalonde, Living with PAH since 2003

Adoption: An Unexpected Blessing

Allison Clarke Wells was diagnosed with PAH in 2017 at age 28. She always wanted to be a mother and was devastated to learn she shouldn't bear children. But only a short time later, Allison and her husband Craig were able to adopt their daughter Penny, a blessing that never would have happened if it hadn't been for her PH.

I always knew I wanted to be a mother. When I pictured what my life would look like, there was never a question that children would be the centre of my world. Every job I've ever had—from babysitting as a teenager, to my eventual career choice of becoming a teacher—has included kids. When Craig and I got married in 2011 we were both still completing university, so we decided to wait until finishing school to try to start a family. We never could have imagined the hurdles we would face before our dream of becoming parents would come true.

In 2017, at the age of 28, I was told I had idiopathic pulmonary arterial hypertension (PAH) and the realization that I would never carry a baby was by far the most devastating part of my diagnosis. I had already gone through two ectopic pregnancies (after several years of trying to conceive), so I had been down the road of infertility, but I still believed that I would eventually have a successful pregnancy. Being diagnosed with PAH took away any and all hope of this dream ever becoming a reality and it absolutely crushed me. It was the hardest time of my life. Not only was I still mourning the two babies I had lost, but I was now also grieving for the babies I believed would have been a part of my future.

When diagnosed, I was cautioned by my doctor that under no circumstance should I ever chance a pregnancy given the severity of my PAH. Not only would I be putting my body under duress for the duration of the pregnancy and delivery, but I would also have to stop taking my medications, as they would not be safe for the baby. This was also not an option since a stoppage in treatment would cause this progressive disease to worsen.

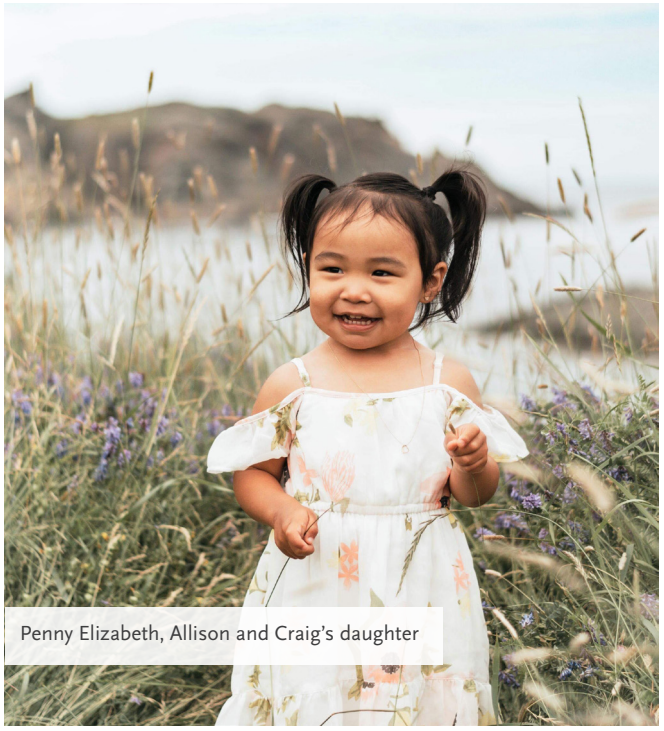
I was dealt this blow after being in the hospital for a month undergoing tests and procedures; dealing with countless doctors and medical jargon I didn't always understand. When I was finally prescribed medications and discharged, I was happy to be able to go home. It was summer and my life pretty much went back to normal—or as normal as it could be—and I rarely spoke about my diagnosis.

> **"Family and friends, who I would see around, would never have known the internal turmoil I was dealing with, facing a future devoid of motherhood."**

I felt empty, like I was "going through the motions" of life without any of the emotion. I played the part—I smiled, I laughed, I went out with friends, I went to family gatherings—but it was all disingenuous. I felt like a shell of the person I once was. I was always sad. PAH took away the joy from my life. Not because of the restrictions it placed on what I was able to do, but because it restricted what I was able to become—a mother.

During this time, the only thing that kept me going was my research on other ways to have a child. I called adoption agencies all over the country; I spoke to social workers and lawyers. My best friend even offered to become our surrogate. We were open to all avenues if it meant that we could start the family that we so desperately wanted. Being told that I would not (or should not) carry a baby to term made me learn so much about other ways to have a child. I read about families that had children through all sorts of miraculous ways and it made me realize that there is no "right" way to start a family. I was making calls, sending emails, reaching out to whomever I could think of who would be able to help us on our journey to start a family.

One day, I messaged a woman I knew who had adopted a little boy, to ask her how she went about starting the process. She told me her son's adoption was a direct placement because she knew the birth mother. The birth mother had chosen my friend and her husband to be the baby's adoptive parents. I was surprised by her story because every social worker and adoption agency I spoke with said we could be waiting for years to be matched with a child, so the likelihood of this process being quick was something I had not heard of. I had that conversation in August of 2017, two months after my PAH diagnosis. On October 1st of that same year, that same friend messaged me and said she was just contacted by her son's biological mother because the biological mother's cousin was pregnant, and she was also looking to place her baby with an adoptive family. My friend wrote "would you be interested?" and at that moment, that unborn baby who was growing inside of a woman I had never met, became mine.



Penny Elizabeth, Allison and Craig's daughter

Allison, Penny, and Craig

Three and half months later our precious girl, Penny Elizabeth, was born and she filled our hearts with more joy than we ever thought possible. She put the broken pieces of my heart back together and made my life make sense again. She saved me. Everything that I thought PH had robbed from me, she gave back.

> **PH took away so much, but it gave me the greatest blessing of my entire life. If it hadn't been for my diagnosis, we may never have approached the idea of adoption and I would have never been given the privilege of getting to be Penny's mother.**

I know that my greatest dream came true because of another woman's greatest loss, and I will always be humbled at the magnitude of the gift Penny's birth mother has given us, while grieving the tragedy of her loss.

Allison and Penny

Becoming a mother after so much grief and heartache has taught me that I need to treasure every moment that I have been given with my daughter. I believe I have gained so much more patience and understanding on my path to motherhood than I would have if it had been an easy road. I take the time to cherish every touch, every snuggle, every laugh, every memory because I am aware of how close I was to never experiencing the joy that comes with being the centre of this tiny human's world. She is my everything, my purpose, my reason for being. She has allowed me to become more than just a person with a rare disease. She has given me a new title—"Mommy"—a title that I will never tire of hearing.

> **While I do wish that I was healthy—that I could run without tiring or go hiking without needing to take a break every few minutes—I am thankful that PH opened my eyes to the different possibilities to create a family and I'll forever be thankful that, in a way, having this disease led me to my daughter. I'm grateful that I see the gift of this life I've been given in a different light and I appreciate every day that I get to spend with the people I love.**

I thank God that he answered my prayers and chose me to be Penny's mother and I promise I'll do my very best to be the mother that she deserves.

Contributed by: Allison Clarke Wells, Living with PAH since 2017

Surrogacy: It Takes a Village

Jane Sernoskie was diagnosed with idiopathic PAH at the age of 26 in 2016. After exploring their options, Jane and her husband Craig decided to work with a surrogate to build their family. They are currently looking forward to starting the IVF part of their journey in early 2021. While at times they find the waiting process difficult, they remain hopeful that this year their perseverance will pay off.

November 2016 was a life altering month for my husband Craig and myself. I was only 26-years-old and was diagnosed with idiopathic pulmonary arterial hypertension (PAH). For some people it can take years to get a diagnosis, but I was diagnosed within minutes of a doctor seeing me. In many ways I am one of the lucky ones because they caught it so quickly. This has allowed me to receive treatments sooner than later and therefore has hopefully slowed down the progression of the disease or halted it for now. I received and continue to receive extraordinary care from my medical team—Dr. Contreras and Advanced Practice Nurse Carolyn Doyle-Cox. They have helped me every step of the way, most importantly by checking in on my mental wellbeing. I truly can't say enough good things about the high level of care I receive from them and other staff at the Ottawa Heart Institute and General Hospital.

Though I was lucky in some ways, I felt very unlucky in other aspects. One of the most shocking parts of my PH diagnosis was learning that it is not recommended to have children. We learnt that carrying a baby would put a lot of pressure on my already strained body and could possibly lead to mine or the baby's death. Craig convinced me it wasn't worth the risk. We talked with my care team about alternative options for growing our family, such as adoption or surrogacy. Ultimately, they told me that they would support us with whatever decision we made, but wanted us to have all the information first. For instance, we talked about the fact that raising a child is extremely exhausting. As a kindergarten teacher, this did not come as a surprise, but it was alarming to hear that raising a child (via adoption or surrogacy) could be very tough on me, more so than someone without my lung condition.

I was devastated to hear this news. Though Craig and I had both envisioned having our own children our entire lives, Craig seemed to work through this shocking news a lot smoother than myself, more easily accepting the alternative routes to building a family. However, I realized I needed some help to get over this speed bump in my life. During an appointment with my GP for a completely unrelated matter, she asked me how I was doing with my diagnosis and I unexpectedly broke down in tears. I was shocked with my reaction because I was trying to be the "strong" one. I saw others around me who shed tears, but I didn't want anyone to see me cry (I now know that crying doesn't mean you aren't strong; it is okay to cry and still be a strong person). My doctor saw these tears and, listening to my story, recommended that I talk to someone about all of my feelings. I thought about it and agreed that I could benefit from some therapy to help me grieve my old normal life.

The psychologist and I met once a month, and I picked up some useful tools that have helped me become more accepting of showing my emotions. One important suggestion she made sounds so simple, but is so hard sometimes for me to do: "be in the moment". I have a tendency to interrupt any sad emotions that arise and redirect myself to think of happier things. She encouraged me to let my emotions out and to nourish my emotional side. She challenged me to create a kindness list and, after I had a good cry, to pick something from that list to do (e.g. snuggle our dog Penny, take a shower, go out for dinner, etc.). I took her suggestions, tried them out, and can see that her ideas were just what I needed.

> **"I needed to purge myself of the grief I was experiencing. These suggestions have had a lasting impact on me, and every now and then I need to remind myself to let my emotions come freely. The combination of the tips from my therapist, coupled with the passage of time and sessions with the Ottawa support group, marked the end of my grieving period. Although, every now and then I still have a cry if that is what my body needs. Fortunately, my acceptance phase followed quickly after my grieving process."**

During my acceptance phase it was like someone lit a match underneath me, as I became extremely driven to continue living my life. I felt like just because I have PAH it doesn't mean I still can't achieve my goals. I may just need to do it in another way. Craig and I began to plan our lives together; we got engaged, bought a house, and got married, all in 2017. We talked it out and together we thought adoption would still be a great option to help grow our family. We considered the fact that raising children is a fulfilling and exhausting journey, and that I may have to sacrifice some of my career aspirations, but it is a sacrifice we are willing to make. So, in the Spring of 2017, "Project Baby" was born.

After doing some research and connecting with other parents who have adopted, including some with PH, we felt like we could make it work too. We then threw ourselves into applying at the Children's Aid Society (CAS) in Ottawa to adopt a baby. Our application was accepted in the Summer of 2017. Months and years passed, and we didn't hear anything from CAS. We understand that the number of babies available for adoption is very slim, so we re-evaluated our plan and began to do some more research into surrogacy.

Meanwhile, my brother and his wife had a baby girl in 2018 who was born on my birthday! We were overcome with adoration and love for her. It fueled us to push on with our journey to grow our family.

Jane and her husband Craig

By January 2019, we shifted our focus to connecting with the Ottawa Fertility Centre to get more information about surrogacy. They told us in order to move forward the first step was to find a surrogate. We opted for the independent surrogacy route, which meant it was up to Craig and myself to find a surrogate. We turned to Facebook and posted a "Wanted: Surrogate" advertisement. I cross-posted it on some surrogacy Facebook pages too. Some of our friends and family helped us by sharing my post on their pages. In a miraculously short period of time we were blessed to find someone that wanted to help us out of the goodness of her own heart.

After several deep conversations with the surrogate, we discovered we were a good match for each other and agreed to continue on this journey together. We went back to the clinic for more advice on how to move forward. After some preliminary tests, in September 2019, we were placed on a waiting list to receive in vitro fertilization (IVF) that would be covered by OHIP (public insurance). It takes roughly 12-14 months on the wait list; however, due to the rise and spread of COVID-19, our clinic was forced to shut and freeze that process for three months. After months of anticipation, in October 2020, our turn came up and we were sent for a battery of tests. The IVF process is now set to occur in the early months of 2021. We remain hopeful that this surrogacy journey will work out and that we will have a baby. We also know there is a chance it may not and that we may need to go back to the adoption path, looking into private adoption agencies and/or expanding our adoption criteria with CAS.

At times, we have found this waiting process to be very difficult, but we believe that one day we will be rewarded for our patience and perseverance, and we will have the privilege of holding our baby in our arms.

Lastly, we'd like to take this opportunity to acknowledge some important people who have been a part of this journey with us.

> **Many people have heard the phrase, 'It takes a village to raise a child', but we are now becoming more familiar with another turn of phrase: 'It takes a village to *have* a child'.**

We would like to extend our deepest heartfelt gratitude to our amazing 'village' made up of our family, friends, care team, and—most importantly our surrogate friend—because without them none of this would be possible. Thanks a million, your support means the world to us!

Contributed by: Jane Sernoskie, Living with PAH since 2016

PH Community Quotes:

What impact has pulmonary hypertension had on your family planning?

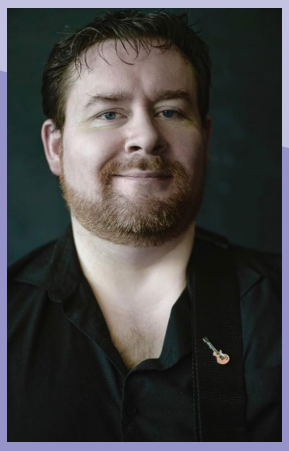
PH: pulmonary hypertension
PAH: pulmonary arterial hypertension
CTEPH: chronic thromboembolic pulmonary hypertension

I have had such a range of emotions on this topic. I have gone from not caring/not wanting a family to having to grieve the loss of something I'm not sure I ever wanted due to society putting so much pressure on women to feel as though they need to be mothers to be fulfilled in life. I am currently in a stage when I have accepted and am embracing my role as the fun aunt. But if history plays out again, chances are there will be another grieving period to look forward to.
—Allison Cain, Living with PAH since 2011, Victoria, BC

Since my CTEPH diagnosis, I have been told by countless high-risk pregnancy and fertility specialists that carrying a child would be life-threatening, extracting my eggs for surrogacy is too dangerous, and that, in any event, my CTEPH medication certainly causes birth defects. There are no words to adequately convey the hopelessness and heartbreak that I've suffered from knowing that motherhood (in the traditional sense) will not form part of my future. And every time I see a pregnant woman or a newborn baby or receive another baby shower invite, my heart breaks a little more. That should have been me.
—Emily Pinckard, Living with CTEPH since 2017, Oakville, ON

The first year after being diagnosed, in 2016, it was difficult for me to plan time for studying and for spending with my young family. Today, after four years of remission from an open-heart surgery that caused my PAH, appointments are fewer and far between. I am fortunate today to lead a virtually normal life as my symptoms have all disappeared. As a mother of now two children and as a registered nurse, I can plan a future with my children and pursue a career helping people, thanks to my great experience with PH.
—Jessica Léonard, Living with PAH since 2016, Trois-Rivières, QC

At diagnosis, I was told I can't "have" children, which to me isn't true. I may not be able to carry a child, but I most certainly can have children. PH has tried to take a lot away from me, has given me a lot of challenges to face, and has dictated a lot of what I can and can't do, but it will not dictate whether or not I can have children in the future. The gift of life comes in many forms. Life is always how you look at it. I may have PH, but PH does not have me.
—Brinley Marks, Age 15, Living with PAH since 2020, Sherwood Park, AB



For me, being diagnosed with PH made my decision regarding children very easy. I was 39 and childless. With the diagnosis came all the usual discussion about life expectancy. I was told it would take 5-10 years before a heart-lung transplantation. I thought it would be selfish and unfair to bring another person into this world knowing that I may not see their 10th birthday, so I opted for a vasectomy. It was the right choice for me.

—Jay Scraba, Living with PAH since 2018, Calgary, AB

When I was diagnosed in 2015, at the age of 36, I was told that I could no longer have children. However, since my diagnosis, the greatest family challenges have been caused by the pandemic. My family has had to make enormous compromises to protect me from possible infection. Since classes began, my daughter has had to live in her room and wear a mask at home. I find it very difficult not to be able to get close to my child and hold her in my arms. For vulnerable people, these moments show us the importance of the people around us.

—Stéphanie Théorêt, Living with PAH since 2015, Candiac, QC

Being diagnosed with PH has impacted greatly on my family planning because I was at an age and stage in my life where I was moving up in my career and I wanted to be married and start a family soon. My world turned upside down the day the doctors said that it is fatal for me to conceive. All I could think of was that no life partner will ever want any relationship with me knowing that I have this life-threatening condition. Attending weddings and baby showers breaks my heart all the time. PH shattered mine and my family's dreams.

—Tasha Gurha, Living with PAH since 2016, Toronto, ON

After a lung transplant, there are so many risk factors involved in starting a family that we thought it best to stay the course and not have children. PH totally flipped my world upside down and forced me to make decisions that I never thought I would ever have to make. But it's also taught me that my life can be incredibly full of laughter, happiness, and joy without children. In fact, I've discovered that being cool Aunt Tina is quite spectacular! It's my favourite job in the whole wide world.

—Tina Giroux-Proulx, Diagnosed with PH in 2003, Received lung transplant in 2015, Ottawa, ON

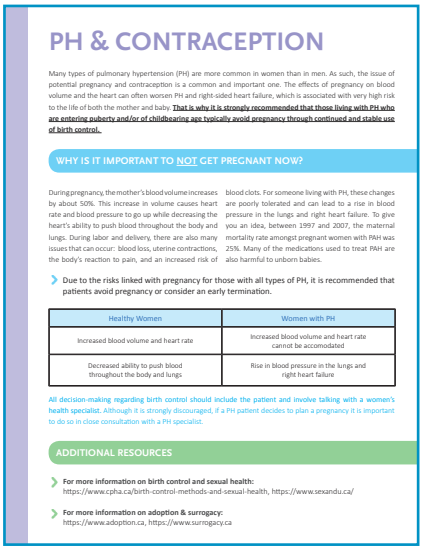
Family Planning Resources

There are a lot of resources available to assist people in making choices about having a family. PH may lead you on an unexpected path, but in the end only you can decide what choice is right for you.

Here are a few important resources to help you make sense of your options.

PH & Contraception

The effects of pregnancy are associated with high risks to the life of both mother and baby, which makes contraception an important issue for those living with PH who are entering puberty or are of childbearing years. Learn more from PHA Canada's downloadable information sheet on PH & Contraception.



PH & CONTRACEPTION

Many types of pulmonary hypertension (PH) are more common in women than in men. As such, the issue of potential pregnancy and contraception is a common and important one. The effects of pregnancy on blood volume and the heart can often worsen PH and right-sided heart failure, which is associated with very high risk to the life of both the mother and baby. **That is why it is strongly recommended that those living with PH who are entering puberty and/or of childbearing age typically avoid pregnancy through continued and stable use of birth control.**

WHY IS IT IMPORTANT TO NOT GET PREGNANT NOW?

During pregnancy, the mother's blood volume increases by about 50%. This increase in volume causes heart rate and blood pressure to go up while decreasing the heart's ability to push blood throughout the body and lungs. During labor and delivery, there are also many issues that can occur: blood loss, uterine contractions, the body's reaction to pain, and an increased risk of blood clots. For someone living with PH, these changes are poorly tolerated and can lead to a rise in blood pressure in the lungs and right heart failure. To give you an idea, between 1997 and 2007, the maternal mortality rate amongst pregnant women with PAH was 25%. Many of the medications used to treat PAH are also harmful to unborn babies.

- Due to the risks linked with pregnancy for those with all types of PH, it is recommended that patients avoid pregnancy or consider an early termination.

Healthy Women	Women with PH
Increased blood volume and heart rate	Increased blood volume and heart rate cannot be accomodated
Decreased ability to push blood throughout the body and lungs	Rise in blood pressure in the lungs and right heart failure

All decision-making regarding birth control should include the patient and involve talking with a women's health specialist. Although it is strongly discouraged, if a PH patient decides to plan a pregnancy it is important to do so in close consultation with a PH specialist.

ADDITIONAL RESOURCES

- **For more information on birth control and sexual health:** https://www.cpha.ca/birth-control-methods-and-sexual-health, https://www.sexandu.ca/
- **For more information on adoption & surrogacy:** https://www.adoption.ca, https://www.surrogacy.ca

phacanada.ca/contraception

The Pulmonary Hypertension Association's Scientific Leadership Council issued a consensus statement on "Birth control and hormonal therapy in pulmonary arterial hypertension" in 2008. The statement discusses preferred birth control methods for women with PAH and provides guidelines that should be discussed with your personal medical team.

phacanada.ca/ConsensusStatement

PH & Parenthood

The stories shared in this issue of Connections are a powerful reminder that there are many ways to build a family. For more information on adoption or surrogacy in Canada, check out:

Adoption Council of Canada
Conseil d'adoption du Canada

adoption.ca

SURROGACY IN CANADA ONLINE

surrogacy.ca

If you have PH and are considering becoming pregnant, it is strongly recommended you consult with your PH team as soon as possible.

Contributed by: PHA Canada

Research Corner

This section focuses on Canadian research and progress towards new therapies in the hope of one day finding a cure for PH.

> **The goal of this research is to improve clinical studies that will evaluate future treatments for PAH so that they meet your needs.**

Researchers at Laval University Identify a New Drug that May Help Treat PAH

When thinking about your health, you would think it was a good thing for cells to be surviving and even thriving. But as doctors have found, in the case of pulmonary arterial hypertension (PAH), that's not always the case.

In PAH, the cells that form the pulmonary arteries—called pulmonary artery smooth muscle cells—divide and multiply rapidly, taking up space inside the arteries and obstructing circulation. Many PAH treatments aim to prevent uncontrolled multiplying of these cells, sometimes even killing them. "It's not intuitive that we have to kill some cells in the body, but the tissue structure is regulated by the number of cells and you can't have cells that keep dividing indefinitely," says Géraldine Vitry, a PhD student under the supervision of Dr. Roxane Paulin, who is part of the Pulmonary Hypertension Research Group at Laval University in Québec, QC. Vitry received the *PHA Canada Paroian Family PH Research Scholarship* in 2019.

Vitry recently led a study to search for new clues about the molecular causes of PAH and to identify new treatment targets. She examined the proteins present in samples from people with PAH and found higher levels of a protein called nudix hydrolase 1 (NUDT1), which helps cells survive and grow under stressful circumstances, such as in PAH.

Interestingly, NUDT1 has already been found to be involved in cancer cell growth and researchers have tried developing drugs to target NUDT1 before, but they had bad side effects and weren't suitable to treat PAH. "But in 2018, there was a study in [the scientific journal] *Nature* showing that the drug (S)-crizotinib specifically targeted NUDT1 and showed anti-proliferative effects to inhibit tumors, so we took this drug and we tested it in our study," says Vitry.

Géraldine Vitry

> **"We hypothesized that probably the same pathway implicated in cancer cell proliferation and survival may be activated in pulmonary arterial hypertension, and that's why we are looking at this pathway and trying to test a drug already shown [to be effective] in cancer therapy."**

In her study, she and other scientists tested the drug (S)-crizotinib in two different animal models of PAH by delivering it directly to the animals' lungs, similar to how medication is administered using an inhaler in asthma, says Vitry.

She found that rats with PAH treated with the drug had significantly better cardiac outputs and lower pressures in their pulmonary arteries than rats that did not receive the drug. Vitry also found that the pulmonary artery walls of rats treated with (S)-crizotinib were not as thick, allowing more blood flow and reducing pulmonary pressures.

When it comes to (S)-crizotinib, a lot of the groundwork in clinical trials is already established. Vitry says researchers need to conduct more studies using pre-clinical animal models of PAH to investigate whether (S)-crizotinib treatment may also help with other aspects of PAH, such as improving muscle metabolism.

"The next step is clinical trials. There have been many ongoing studies, especially in the last 20 years, that showed that we can slow disease progression using some disease pathway, but there are many studies that don't succeed in clinical trials because humans are different than animals," says Vitry. "Also, not all patients will have high levels of NUDT1 and have the same molecular signatures. So, it may not help every patient, but we need to make the effort to look at different patients' molecular makeup to see which treatments would be best for them."

Vitry explains that because there is no evidence that the high levels of the NUDT1 protein (which causes cell proliferation) are due to a genetic mutation of its gene, it's possible that in different people with PAH there will be different reasons for the increase in NUDT1. So, using a drug that directly affects NUDT1, such as (S)-crizotinib, can decrease NUDT1 protein levels no matter what the cause, potentially benefiting a wide range of people with PAH.

"You don't always see the human side when you're working so much—we look for solutions, but aren't affected by [PAH]," added Vitry while thinking about the time a mother of a child with PAH visited the PH research lab at Laval. "That reminded me that there is an urgency in this disease for adults, but also children. There are people that need help."

Even if (S)-crizotinib doesn't pass clinical trials, Vitry's research is an important part of understanding what happens in pulmonary artery smooth muscle cells in PAH and another step toward finding new treatments.

Contributed by: Miriam Bergeret, Living with PAH since 2017, PHA Canada Knowledge Translator

Miriam Bergeret

Share Your Expectations for Current and Future Medications to Treat Pulmonary Arterial Hypertension

Participate in this national survey at phacanada.ca/TreatmentSurvey.

Over the past two decades, several new treatments for PAH became available. The development of new drugs is a long and complex process that requires several phases of study. Ultimately, phase three studies are performed following the recruitment of a large number of subjects, who are drawn at random, and comparing the effectiveness of the new treatment with a placebo (tablets without the active ingredient) to achieve a specific goal determined before the start of the study.

Treatment goals can obviously vary from disease to disease, but also depend on the treatment setting or individual person.

With the introduction of the first treatments for pulmonary arterial hypertension (PAH) in the late 1990s, the main goal was obviously to increase the survival of people suffering from this fatal disease. Given the relatively strong link between the ability to exercise, quality of life, and long-term survival in people with PAH, studies in the early 2000s focused on evaluating the effect of new therapies on the ability to exercise in people with PAH who were not receiving basic treatment.

Unlike participants in the studies conducted in the 2000s, people with PAH now have access to treatment. Since their condition is already improved by an initial treatment, it is increasingly more difficult to demonstrate that the addition of another treatment further improves the ability to exercise. It also seems obvious that a six-minute walk test does not reflect the overall condition of a person with PAH.

In fact, the goal is not only to maintain or improve the ability to exercise, but also to prevent adverse events such as hospitalizations or decreased performance on the six-minute walk test.

In recent years, studies have aimed to demonstrate that adding a new treatment can prevent a series of adverse events. At the end of the study, it is thus analyzed whether these events occurred less often, in fewer people, or later in the group receiving the new treatment compared to the group receiving the placebo.

However, this list of events is defined by the researchers responsible for the study rather than people who suffer from the disease, and little information is available on what is really important to them. While the relevance of preventing death is obvious to everyone, the importance given to preventing hospitalizations or a slight decrease in performance on the six-minute walk test may differ between a researcher, a person with PAH, and his or her caregiver. Even among people with PAH, this importance can fluctuate depending on age, values, stage of disease, etc.

In order to clarify the importance that people with PAH, their caregivers, and their medical team place on preventing each of these events, we recently launched a pan-Canadian study on the subject.

> **The goal of this research is to improve clinical studies that will evaluate future treatments for PAH so that they meet your needs.**

The project is supported by the Pulmonary Hypertension Association of Canada (PHA Canada), the Pulmonary Arterial Hypertension Foundation of Quebec (Fondation HTAPQ), and the Institut Universitaire de Cardiologie et de Pneumologie de Québec (IUCPQ).

This new study follows the principle of "patient-centered research", as reflected in the most recent *Pulmonary Hypertension Priority Setting Partnership* survey (phpsp.ca). This approach to health research takes into account the perceptions, concerns, and wishes of people with PAH and their caregivers to ensure that the studies conducted meet their needs.

This project begins with a national survey which will take place in the winter of 2020-2021. Participation is voluntary and anonymous. The questionnaire, available in English and French, will ask you some basic information about your situation and will invite you to indicate the importance you give to the prevention of a series of events (hospital admission, starting a new treatment, etc.). We will then examine:

1. whether the answers differ between people affected with PAH, their caregivers, and their medical team;
2. the factors that seem to influence the results (e.g., age, severity of symptoms, etc.);
3. how previous clinical studies actually aimed to prevent events that were considered important by PAH patients, their caregivers, and their medical team.

If you are a person with PAH or are caring for someone with PAH, your answers are important to us.

Pulmonary Hypertension Research Group

Contributed by: Steeve Provencher, MD, MSc, FRCPC, Respirologist in charge of the Pulmonary Hypertension Program, Québec Heart and Lung Institute, Université Laval

PHA Canada Awards $100,000 in Research Scholarships Since 2016

PHA Canada is proud to announce the recipient of our 10th research scholarship award. The $10,000 scholarship marks a total of $100,000 in research scholarships awarded since 2016. PHA Canada currently supports up-and-coming Canadian research through two named PH research scholarship funds: the *Paroian Family PH Research Scholarship* and the *Mohammed Family PH Research Scholarship*. We are grateful for the continued support of the fundraisers and donors who make these scholarship funds possible.

We are pleased to announce that the fifth recipient of the *PHA Canada Paroian Family PH Research Scholarship* is Austin Read, MSc Candidate, Masters of Translational Medicine, at Queen's University's Faculty of Health Sciences, Department of Medicine, in Kingston (ON).

Austin Read, MSc Candidate
Queen's University, Department of Medicine (Kingston, ON)

Under the supervision of:
Dr. Stephen L. Archer, Head of the Department of Medicine
Queen's University, Faculty of Health Science (Kingston, ON)

Prior to attending Queen's University, Austin Read completed his Bachelor of Science, with a specialization in Chemical Biology, at McMaster University in 2019. He is now in the second year of his graduate studies in translational medicine.

He is currently investigating the mechanisms that underlie fetal-to-newborn cardiovascular changes, specifically looking at the role of specific protein complexes in how cells sense changes in oxygen levels at birth. In Read's project, there is also potential of identifying new therapeutic targets to treat persistent pulmonary hypertension of the newborn (PPHN).

Austin Read's Project: ***Mechanisms of fetal oxygen sensing and the role of the electron transport chain in the pulmonary artery and ductus arteriosus***
Research team: Austin Read, Queen's University (Kingston, ON), Dr. Archer's Lab

With the first breath, a newborn's circulatory system must quickly adapt to obtaining oxygen through respiration, rather than through their mother's circulation. In the fetus, the heart is designed to allow oxygenated blood coming from the placenta to bypass the developing lungs and be re-directed to the body's circulation by way of a large fetal vessel called the patent ductus arteriosus (DA). When we take our first breath at birth, a rise in airway oxygen causes the pulmonary arteries to expand and the DA to constrict and eventually close, initiating the transition of the circulation from fetal to newborn patterns. When the DA fails to constrict, this results in persistent pulmonary hypertension of the newborn (PPHN), a major cause of congenital heart disease in newborns.

Both pulmonary artery expansion and DA constriction are dependent on the ability of these tissues to sense and respond to oxygen in their surrounding environment. Thus, it is likely that disorders of oxygen sensing contribute to PPHN.

Read's proposed project aims to identify how cells of the fetal pulmonary arteries and DA sense and respond to oxygen in their environment, in order to identify new molecular targets to treat PPHN.

Contributed by: PHA Canada

Canadian PH Community Survey

"The survey results will allow us to better understand the Canadian PH community and will also give us an opportunity to focus on areas that most need improvement."
—PHA Canada Board Chair Nicole Dempsey,
Living with PAH since 2013

Share your experience with PH!

Coming this Spring

Empower the Canadian PH community:
phacanada.ca/Survey

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Contribute

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

Send your contributions to: info@phacanada.ca

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.

The Pulse

PHA Canada's monthly newsletter

The Pulse will keep you updated on what's happening in the world of PH in Canada and beyond.

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- Community event announcements
- Research news from Canada & around the world
- Low-sodium Recipe of the Month
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