

# PNH Review

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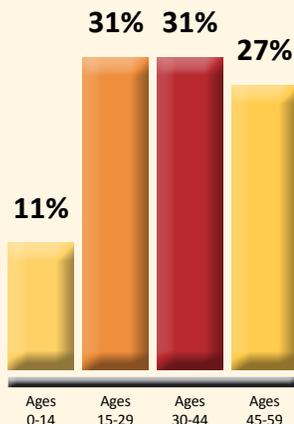
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## What You're Telling Us

Our most recent poll question on [www.pnhca.org](http://www.pnhca.org) asked respondents how old they were when they were diagnosed with PNH. 31% of patients were diagnosed between the ages of 30 and 44, and 31% were diagnosed between the ages of 15 and 29. Finally, 27% of respondents were diagnosed between the ages of 45-59, and 11% were diagnosed between the ages of 0-14.

We appreciate your responses to our survey questions, and we look forward to sharing our next poll question and results with you soon! Your participation in our polls is helping us to better understand the PNH community in Canada, so please cast your vote in the next poll question at [pnhca.org](http://pnhca.org)!



## Where Are They Now?

In March of 2013, Vancouver's Tracy Murray began to experience severe fatigue, nausea, and abdominal pain and swelling. Concerned about these symptoms, she went to numerous walk-in clinics and was sent for blood tests to find out what was wrong. By the end of May 2013, Tracy went to the hospital and was immediately admitted to the emergency room, and doctors released fluids from her stomach through a pump to ease her pain. Over the following days, she underwent a series of tests, x-rays and scans, and doctors discovered a blood clot in her liver and rushed Tracy to Vancouver General Hospital.



Tracy was diagnosed with Budd-Chiari syndrome, a complication that many PNH patients experience. In Budd-Chiari, blockages occur in the hepatic veins, which drain the liver. Tracy was put on warfarin (a blood thinner) and had a stent surgically inserted into her hepatic vein to prevent additional clots from forming. Shortly after this episode, and after undergoing numerous tests, Tracy received her diagnosis of PNH (paroxysmal nocturnal hemoglobinuria), which explained her recent health issues.

By July 2013 – only two weeks after returning home from the hospital - Tracy was readmitted to hospital with another severe clot and swelling. She underwent a second operation to widen the stent in her hepatic vein.

Thanks to private medical coverage through her employer, Tracy was able to begin treatment with Soliris, the life-saving medication proven to return patients' lives and health to normal. Tracy received her first life-changing infusion on August 4, 2013 – and hasn't looked back.

Now married to a loving, supportive husband and working as a travel agent, she says her energy levels have quadrupled. She no longer experiences the extreme exhaustion or the constant worry that a blood clot could instantly end her life. "I can finally function again," she says. Because of the amount of damage and the number of surgeries her body endured, she admits that her recovery has taken longer than is common. "I certainly consider myself lucky, but it did take me a while to get 'normal.'"

Today, Tracy is able to enjoy a fulfilling personal and professional life, and loves to educate others about PNH and serve as a role model to other patients. While she needs one more surgery to address a hernia, she does worry about her ability to access Soliris in the future.

Tracy continues to receive Soliris through private insurance through her employer, but she knows that this coverage may not last forever. "I worry about what might happen to me if my employment or funding situation were to change," she says, "I hope that I won't ever have to face losing access to this life-saving medication."

Like all PNH patients in Canada, Tracy needs the assurance that the B.C. government will make this treatment accessible to her should she ever require it through public funding. "While Soliris is available through government funding, if the time comes, I need to be sure that I would seamlessly transition to accessing the treatment through this means."

Tracy is grateful for the funding support B.C. Pharmacare has provided to PNH patients since Soliris first became accessible in July, 2011. She is calling on the province to maintain its commitment to PNH patients so that no one is left with the stress or anxiety that accompanies any uncertainty around accessing this transformative treatment.

**EDITOR'S NOTE:** New patient stories needed! If you have been recently diagnosed with PNH and would like to share your story with the Canadian PNH community, we'd love to hear from you. Please email us at [contact@pnhca.org](mailto:contact@pnhca.org) today.

# What's Happening?

Calling all PNH patients and caregivers!

You're invited

The Canadian Association of PNH Patients is pleased to invite patients and families from across Canada to attend our next meeting in Toronto, Ontario on **April 11, 2015**. In October 2014, we held a similar meeting in Vancouver for families in B.C., and for this upcoming meeting, we would like to extend an invitation to those in other parts of the country. To see the official invitation and to download the registration form, please visit [our website](#).

The event will be held at the Novotel North York hotel from 9 a.m. to 3 p.m., with breakfast and lunch provided. There will also be a welcome dinner for families on the Friday evening. PNH expert Dr. Richard Wells of the Sunnybrook Health Sciences Centre, who is also chair of the Canadian PNH Network, will deliver a clinical presentation on PNH at the meeting.

We look forward to meeting new families from across Canada at this event – as well as seeing some familiar faces! Please contact [zilla@pnhca.org](mailto:zilla@pnhca.org) with any questions, and to see if you qualify for a travel subsidy. The deadline for registration is **Friday, March 27, 2015**.

## ***Listening To You: A Program To Understand Your Journey***

In January, the Canadian Association of PNH Patients invited members of the Canadian PNH community to participate in “*Listening To You: A Program To Understand Your Journey!*” The project represented a new platform to enable patients to share their experiences with regards to PNH and its treatment.

The Association would like to thank those individuals who participated in this important research project! We also expect to have results ready to share at the upcoming patient and caregiver meeting in Toronto on April 11. Stay tuned!



## ask the expert

We've added new questions to the **Ask the Expert** feature on the PNH website – [click here](#) to see them!

To share your tips for coping with PNH and its symptoms, send your suggestions to [contact@pnhca.org](mailto:contact@pnhca.org)

### **I am on Soliris. Is it recommended that I take antibiotics on an ongoing basis? If so, why?**

Soliris works by blocking the activation of the terminal complement pathway. This protects PNH red blood cells from destruction, and has a huge beneficial effect for PNH patients. However, Soliris also increases patients' vulnerability to infection with *Neisseria meningitidis*, a type of bacteria that causes meningitis. All patients taking Soliris must be vaccinated against *Neisseria meningitidis*, but until recently, none of the vaccines available provided protection against the most common “serotype” of this bacteria. This means that even individuals who had been vaccinated could still get this infection. For this reason, many PNH physicians recommended continuous antibiotic therapy to provide additional protection against this dangerous infection.

### **I understand there is a new meningitis vaccine available. Who should have it and when?**

Health Canada recently approved a new meningitis vaccine, Bexsero, for use in Canada. This vaccine covers the most common serotype of *Neisseria meningitidis*, which was missed by previous vaccines. Although Bexsero has been tested most extensively in children, most PNH physicians recommend it for adults with PNH because of the high risk of *Neisseria meningitidis* infection in patients receiving Soliris. Bexsero is given as two injections several weeks apart. Currently, it is recommended that all patients starting Soliris have the first Bexsero vaccination before the first dose of Soliris. Patients already receiving Soliris should have Bexsero on the day of a Soliris infusion. The vaccine is always given on the day of a Soliris infusion because it activates the complement system and may provoke an increase in hemolysis.

This information was prepared by the Canadian Association of PNH Patients and reviewed by: Dr. Richard A. Wells MD, D.Phil., FRCP(C) Hematologist, Sunnybrook Health Sciences Centre and assistant professor, Department of Medical Biophysics, University of Toronto.

Questions, comments or ideas for the next issue? Please send us an email at [contact@pnhca.org](mailto:contact@pnhca.org).