



# CanPKU News

Issue No. 12

Canadian PKU & Allied Disorders Inc. Newsletter

June 2013

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## Dear Reader,

With a multitude of events happening across the country over the last few months, it's really been a challenge getting everything into the newsletter this issue.

I want to keep this section short and just briefly mention that although all of our provincial PKU Events have now been held, as well as our first ever National Walkathon, due to space restrictions we have chosen to highlight a few of our events in the next issue rather than this one. Please don't be offended, we merely want to ensure every article gets a fair highlight!

We felt it important to inform everyone of the soon to be released ACMG guidelines, as these could have a significant impact in clinical practice across the country. As such, this is our headline article.

Finally, we have also included tributes in memory of two people special to the PKU community.

Best,

Ruth Appanah

Editor

CanPKU and Allied Disorders Inc. Newsletter



Innovation in Nutrition

### PKU Profile\*

### Brett Hoover



Brett Hoover was born in Calgary in 1999 and was diagnosed with PKU at Alberta Childrens Hospital when he was three weeks old.

Brett now lives in Saskatoon with his parents, Jodi and Tim, his older brother Blake, younger brother Benjamin and his little sister, Brooklyn. He has a close relationship with both sets of grandparents and with his many aunts, uncles and cousins who live nearby.

Brett sometimes struggles with

### New Guidelines for PKU Treatment about to be released

Here is a preview of new guidelines being finalized by the American College of Medical Genetics and Genomics (ACMG). These guidelines will prompt questions and potential changes in the goals for PKU treatment and raise issues about the differences in practices among clinics.

The committee writing the PKU guidelines was chaired by Dr. Jerry Vockley of Pittsburgh and included two Canadian specialists, Dr. John Mitchell and Dr. Nancy Braverman, both of Montreal. Dr. Vockley has spoken at patient/family events across Canada since 2009 in Halifax, Moncton, Montreal, Toronto (Alliston), Red Deer and Vancouver. He is scheduled for a patient/family event in Edmonton in October. Another committee member was Dr. Barbara Burton of Chicago who has spoken at patient/family events in Moncton, Montreal, Toronto (Alliston) and Vancouver.

Two major conferences sponsored by the US National Institutes for Health (NIH) held in 2000 and 2012 reviewed the state of knowledge of PKU but there have been are no generally accepted recommendations for therapy. Nicole Pallone and John Adams attended the NIH conference held last year.

#### ACMG draft Recommendations:

1. The goal of treatment should be to maintain Phe levels between 120 and 360  $\mu\text{mol/L}$  (2-6  $\text{mg/dl}$ ) for all patients of all ages.
2. Treatment should be lifelong for patients with untreated Phe levels of more than 360  $\mu\text{mol/L}$  (6  $\text{mg/dl}$ ).

These recommendations are bound to cause a stir although the use of the guidelines by clinicians is voluntary.

For context, there are 21 clinics across Canada treating PKU patients. A recent survey by the Garrod Association indicated that about half of the clinics which answered the survey said they wait for higher phe levels before they begin treatment. Three clinics said they do not treat until levels reach 600 and others are in between 360 and 600. So the second recommendation could effect what happens in those "high" clinics, especially for newborns or anyone newly diagnosed.

compliance of his PKU diet, but he likes cooking and baking. He is very creative in the kitchen and makes most of his own meals himself.

Brett plays the acoustic and bass guitar, enjoys singing and participates in competitive hip hop dance. He also enjoys performing in various school talent shows and musicals. Brett is very interested in computers and is looking forward to going to high school next year.

Jodi, a Kindergarten teacher, has been advocating for PKU in Saskatchewan for several years. She met with the Minister of Health, Don McMorris, in February, 2009 and is very grateful that the provincial government expanded the formula program and is covering some low protein foods as of January last year. She continues to advocate for access to Kuvan and other medical services and treatment options.

\* Please note that we are always looking for children or adult profiles to highlight with each newsletter. If you would like to contribute your or your child's story please contact the editor!

### **Featured Recipe**

#### **MARIA'S BANANA BREAD\***



125g margarine or butter, unsalted  
110g sugar powder  
210g low protein baking mix (any brand)  
1 1/2 tsp baking powder

The first recommendation could effect the treatment of older children, teens and adults.

Since an international scientific conference in 2000, there has been a widely accepted consensus that Phe levels in younger children with PKU should be kept below 360, although there was disagreement among physicians whether or not to treat hyper-phe patients with levels from 360 to 600.

Many clinics "relax" the goal of treatment for older children, teens and adults. The argument is that asking teens and adults to keep their levels below 360 is so difficult and discouraging that it is counter-productive. However, there is no science to show that levels above 360 are safe at any age and instead we should focus on the need for improved treatments.

The ACMG draft guidelines highlight that:

- Patients who discontinue therapy will likely experience neuropsychological improvements by going back on therapy.
- Patients with late or untreated PKU may benefit from starting therapy

The draft guidelines also recommend:

- any combination of therapies, such as diet and Kuvan, that improve Phe levels is appropriate
- therapies may be combined and should be individualized
- the risk for neurocognitive or psychological symptoms in PKU is related to age of onset of therapy, lifelong Phe levels, and adherence to treatment
- age-specific neuropsychiatric and cognitive testing is necessary to adequately assess clinical needs.
- appropriate intellectual and mental health assessments are an important component of care for individuals affected with PKU
- Treatment-for-life mandates the need to provide coverage for medications and medical foods regardless of age.

**The ACMG draft guidelines are informed by an evidence review undertaken by the US government's Agency for Health Research and Quality.**

This evidence review noted that the probability of IQ in PKU patients being less than 85 increases when Phe levels go above 360 umol/L.

1/4 tsp salt  
1/4 tsp ground nutmeg  
1/4 tsp cinnamon  
200g ripe bananas, mashed  
60ml water

In a large bowl mix sugar powder, baking mix, baking powder, salt, nutmeg and cinnamon. Add melted butter, mashed bananas, water and mix well. Pour the liquid dough into bread baking form and bake in preheated oven at 350F for about 20-30min, until the crust is golden and knife, inserted in the middle comes out clean.

Per recipe  
Calories: 2173 cal  
Protein: 3.86 g  
Phenylalanine: 150mg  
Recipe yield: 5 portions  
Per portion:  
Calories: 434.6 cal  
Protein: 0.8 g  
Phenylalanine: 30 mg

\*From The Wooden Spoon: [click here](#) to see more!

## **Introducing a New Contributor!**

Maria Depenweiller, an adult PKU'er, nutritionist, and professional home economist, has kindly agreed to donate her time in the form of regular recipe and/or nutrition updates for our quarterly newsletter.

Maria is the owner of her own business The Wooden Spoon!

The Wooden Spoon provides services including educational seminars and workshops, recipe development and testing, food writing, and consulting to those in need. Because Maria herself has PKU, she is able to pay special attention to the PKU community. One of the specialties of The Wooden Spoon is the development and testing of low protein

## **2013 Event Series Updates**

Prairies PKU Day was held on Saturday April 26

<sup>th</sup> in Saskatoon, SK. We had wonderful weather and a great turnout of 60 people! The day started off with

a 2 hour presentation by renowned PKU expert, Professor Laurie Bernstein, RD. Local event coordinator Jodi Hoover enlisted the help of many friends and family members and the day went off without a hitch. Attendees enjoyed a catered lunch (both regular and low protein) and many samples from metabolic vendors. After lunch, the kids enjoyed returning to the daycare while the older teens and adults heard about Dr. Robin Casey's experience in the PKU-016 study, and from Nicole Pallone, CanPKU Vice President, about her experience managing her young daughter's PKU with a combination of low protein food, formulas and Kuvan. There was plenty of opportunity to mingle and everyone had an enjoyable day!



Our 2013 Event Series continued with a stop in Halifax, Nova Scotia, for Atlantic PKU Day. We were honoured at this event to have Dr. Kendra Bjoraker, a neuropsychologist, donate her time to CanPKU for the first time. Her presentation was informative, interesting and inspiring - watch for her on future programs! There was a focus on advocacy as many patients in Atlantic Canada struggle with a lack of funding for treatments and for the first time in CanPKU history, the attendees represented more adult PKU patients than child PKU patients! It was great to see so many adult patients taking an active role in their healthcare by attending our event and learning more...

recipes and low protein cooking workshops. This can be particularly useful in teens/adults starting college life or anyone who might need some help with their cooking!

Throughout 2012 Maria ran several great workshops around the Toronto area such as a tea tasting workshop at Richmond Hill community centre and whole grains hands-on cooking session at Davenport Perth Health Centre. Throughout the summer Maria also participates in organizing children's cooking classes at the High Park teaching kitchen.

Check out Maria's low protein food photography on Instagram by following #phenylketonuria and #lowprotein.

For more information on The Wooden Spoon please see:

[www.thewoodenspoon.ca](http://www.thewoodenspoon.ca)  
<http://thewoodenspoonblog.wordpress.com/>

Finally, If you have any questions or wish to book a workshop feel free to contact Maria at [mdepenweiller@gmail.com](mailto:mdepenweiller@gmail.com)



***Because Knowledge Leads to Better Health!***

Stay tuned for recaps of our Quebec and Ontario PKU Days coming in our September 2013 newsletter!

### **CanPKU Penny Drive**

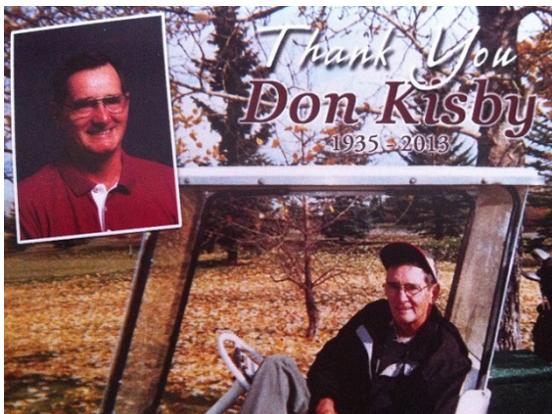
Fundraising is an essential part of the activities of CanPKU, required to support all the advocacy and related tasks that the organization undertakes.

One of the ways you can help? Well, the CanPKU national penny drive is still on-going! Please continue to collect pennies from your friends and family members and donate them to us! Every little bit can make a difference!

For more details on how to help please [click here!](#)

**Your support is greatly appreciated!**

### **Don Kisby Memorial Tribute**



Don Kisby loved life, and enjoyed much of it with his loving wife Gail at his side. He passed away February 21, 2013 after a courageous battle with cancer. He was a plumber by trade, an avid golfer, and a talented wood worker making toys and scale model trucks.

Without children of their own, they devoted time to supporting nieces and nephews and friends alike. Don and Gail were among the first to know when news came that their great nephew Ethan was born in 2001 with a rare genetic disorder called PKU. They were also there to wipe away the tears and support the process of learning to manage a foreign diet and way of living. And proudly they have been there to rejoice in every success and milestone for Ethan.

Ethan is now 12 years old, a bright, athletic, tech loving, regular kid. He has had challenges with formula and diet at times like most kids with PKU, but takes it all in stride. He participated in a Kuvan trial last year and responded well with a decent reduction in phenylalanine levels as well as improvements in ability to focus his attention at school and in sport.

Uncle Don was one of Ethan's greatest supporters in life and in memoriam.

In loving memory of Uncle Don, he together with Gail chose to honor CanPKU by requesting memorial tributes in lieu of flowers, and as a testament to what a kind and generous person Don was, there was an overwhelming response.

### **Advocacy Updates**

CanPKU volunteers spend an extraordinary amount of time advocating across the country for better access to treatments for PKU patients. Our initial campaign launch in BC has been met with some fierce resistance but we have held strong in our belief that BC patients deserve better. It is important to note that while the clinical services and access to formulas are very good in BC, our focus is on the complete lack of access to medically necessary foods and Kuvan. However, we are hopeful that a new Minister and Deputy Minister of Health will

### **NATIONAL CANPKU WALKATHON!**



As mentioned in the last newsletter, 2013 also marks the 1st National CanPKU Walkathon! This was held on **Saturday, June 1st**, with five official venues (and one surprise venue!) across the nation. As expected, the Walkathon was a great way to raise awareness about PKU and our organization. Here are some facts and figures from the event:

**2-4:** the number of hours needed from a local host to organize a CanPKU walkathon venue.

**6:** the number of walkathon locations across Canada (including an unregistered walk in Prince George, BC!)

**107:** the number of walkers who participated in the event on June 1st.

**5,991:** the number of dollars raised for CanPKU!



**We sincerely thank all the organizers for their contributions to a successful event:**

be more responsive to our requests in the coming months.

We are also very active in other jurisdictions. CanPKU was very involved in discussions with the Ontario government as they reviewed Kuvan and ultimately chose to fund it for PKU patients (we do still have some work to do there as the criteria adopted is very strict). We have also begun discussions with the Newfoundland & Labrador Ministry of Health regarding expansion of funded treatments and inclusion of adult patients as beneficiaries of the program. Atlantic Canada also has a unique situation in which nearly a dozen families have been uniformly denied funding of Kuvan by a single insurance company; CanPKU has written to that company on behalf of these patients and families and will continue.

We know there is serious work to be done in every province: quite simply, no province yet has coverage and care that provides access to all the tools in the toolbox for every patient. We have access to some fabulous resources in regards to advocacy training and we are committed to continuing our advocacy work as one aspect of fulfilling our **Mission Statement: to improve the lives of people with PKU and allied disorders and the lives of their families.**

### **Upcoming PKU Summer Camps!!!**

The list below highlights a number of camps scheduled through the summer in 2013 which would be suitable for those on a low-protein diet.

**June 24-29, 2013**  
**Metabolic Camp at Emory University - Atlanta, Georgia**

**July 14-19, 2013**  
**Camp PHEver - Burton, Texas**

The London, Ontario walk was hosted by CanPKU Director, Jennifer Ballagh.

The Kamloops, BC walk was hosted by PKU adult and long-standing CanPKU volunteer, Amanda Cosburn.

The Red Deer, Alberta walk was hosted by PKU Mom, Amy Christenson.



The Sparwood, BC walk was hosted by CanPKU Vice President, Nicole Pallone

The Vancouver, BC walk was hosted by new PKU mom Caylee Auge.

**The 2014 National CanPKU Walkathon will be held on Saturday, May 31st!! Please mark your calendars and please consider hosting a walkathon in your community next year!!**

August 16-18, 2013  
Maryland Alliance of PKU Camp Huber -  
Chestertown, Maryland

August 16-19, 2013

Cavett Kids Foundation Camp - Pryor,  
Oklahoma

August 18-23, 2013 - South Shore YMCA PKU  
Camp - Sandwich, Massachusetts

August 23-26, 2013  
Oregon PKU Family Camp - Antelope,  
Oregon

For more information or to register for any of  
the above camps please check out the NPKUA  
event calendar by [clicking here!](#)

## Become a Member!

New members will receive one free issue of National PKU News, courtesy of Virginia Schuett and all members now receive a 5% discount on all Country Sunrise products ordered from PKU Perspectives! Other benefits include discounts on CanPKU event registration fees, eligibility for CanPKU Travel Scholarships and voting rights at our Annual General Meeting (General Members only).

To become a member go to:  
<http://www.canpku.org/become-a-member>.

Please ensure that you choose the appropriate category when you next renew your membership!

- **General Member (GM):** any person diagnosed with PKU or other allied disorder; any person related to a person with PKU or other allied disorder and whose life is directly affected by the diagnosed disorder (such as a parent, grandparent, legal

## Mourning the passing of a little-known Hero of PKU

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Who was the first doctor, in the world, to treat a PKU patient successfully?

Answer: A modest but distinguished pediatrician named John W. Gerrard, way back in 1951.

Dr. Gerrard moved to Canada in 1955 to be the first head of the new pediatrics department at the new full medical school in Saskatchewan and remained its head until 1971. He continued to treat patients until well into his 80's, as Professor Emeritus of Pediatrics. He passed away at the age of 96 years on March 3, 2013 in his adopted country, province and city of Saskatoon.

Born in 1916 in Zambia, Africa to medical missionary parents, he trained as a doctor at Oxford University, England. Early in his career, he ran a clinic for children with intellectual disabilities (mental retardation) of unknown causes at the Children's Hospital in Birmingham, England, where he had trained in his chosen specialty as a pediatrician.

In that clinic was a 2-year-old girl named [Sheila Jones](#). A medical colleague, Horst Bickel, used a new kind of test and diagnosed the girl as having PKU. Her mother repeatedly asked the clinic what they could do to help her daughter. As a result of that persistent mother, this clinic led by Dr. Gerrard was the first in the world to use a newly-published idea to make the first low-phe formula for PKU treatment. It worked, was duplicated in London, England and changed the worlds of PKU and genetics forever.

In Saskatoon, he taught hundreds of medical students and helped train over a hundred pediatricians. His care, compassion and warmth were exemplary; many a time he and his wife, Betty, opened their home to parents who brought sick children from distant corners but had no means to pay for a hotel room.

In an interview, Dr. Gerrard told me, "I could not make a living in Saskatchewan as a doctor treating PKU as there is less than one PKU birth a year" in that province.

He received the Alan Ross Award from the Canadian Pediatric Society for his contributions to research,

guardian, sibling, spouse or child). Only General Members 18 years of age and older having voting rights.

- **Supportive Member (SM):** any person who supports the vision and mission of CanPKU and is interested in advancing its objectives. Supportive Members have NO voting rights.
- **Professional Member (PM):** any professional (doctors, researchers, nurses, nutritionists, among others) related, interested and/or working on PKU and other genetic and/or metabolic disorders. Professional Members have NO voting rights.
- **Corporate Member (CM):** any corporation interested in helping and supporting CanPKU. Corporate Members have NO voting rights.

**By becoming a member you are showing your support for CanPKU to accomplish its goals, which include:**

- Creating awareness about PKU and other inherited metabolic disorders;
- Providing a supportive community for those living with PKU and other inherited metabolic disorders;
- Increasing opportunities for PKU families and others to attend educational and networking events;
- To improve the lives of people living with PKU and other inherited metabolic disorders;
- Promoting and supporting research; and
- Advocating for increased treatment coverage across Canada.

education, healthcare and advocacy for the health of children in Canada and he was an Officer of the Order of Canada.

Photo: Sept. 11, 2010 Dr. John Gerrard (seated in centre) at CanPKU event, honoured as a Hero of PKU.



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Please feel free to forward this newsletter on to your contacts. CanPKU is always looking for volunteers, members and corporate sponsors. For more information about our organization and other PKU resources, please visit our website at [www.canpku.org](http://www.canpku.org).

Sincerely,  
Canadian PKU & Allied Disorders Inc.



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