

# CanPKU News

Issue No. 11

Canadian PKU & Allied Disorders Inc. Newsletter

March 2013

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### Dear Reader,

Happy Easter (almost) and welcome to the first newsletter of 2013. We have much to update our readers with!

The first of our many event series has been held now - BC PKU day was February 23rd in Vancouver, BC - and was a very well attended and successful event. This was my second time as an attendee and it was certainly a different experience having now had 11 months of being a PKU mom under my belt than the first time around, when our baby girl was only 2 months old!

The next PKU event will be in Saskatchewan, following by many more. Furthermore, in May it will be National PKU Awareness month and immediately afterward, CanPKU is holding a first ever National Walkathon. Please see below for details of all these and more.

I hope you enjoy the issue, and do let me know if you have any suggestions!

Sincerely,

Ruth Appanah

Editor

CanPKU and Allied Disorders Inc. Newsletter





Innovation in Nutrition

## **PKU Profile\***

## John Adams (Jr.)

Most of you will surely recognize the name of John Adams. This newsletter profile is on John's son, also named John!

Their story begins, like most others, with that memorable phone call from the Dr's office telling them they needed to attend the PKU Clinic the following day. That was the first they had heard of PKU, and was before the internet, so John and Marita looked the term up in some reference books. Their first memory was of reading about "profound, irreversible mental retardation."

Today, says John, "our son, John, is today a mature young man, a university graduate, living independently and pursuing his career goals," but "there have been challenges along the way."

# **2013 Event Series and 1st National CanPKU**Walkathon

Our 2013 Education & PKU Community Events are now all confirmed. As mentioned previously, these events are a wonderful way to connect with other PKU families and to learn more about current treatments and research projects.

Saturday, April 27th: Saskatoon, Saskatchewan Saturday, May 4th: Halifax, Nova Scotia Sunday, May 5th: St. John's, Newfoundland Sunday, June 2nd: Sherbrooke, Quebec Saturday, June 8th: Toronto, Ontario

Remember that we try to make travel scholarships available for every event, so don't let financial worries stop you from attending!

If you are interested, please email Nicole Pallone at <a href="micole.pallone@canpku.org">nicole.pallone@canpku.org</a> or call 250-910-6426.

We are passionate about increasing awareness and education about PKU and hope to see you at the events in your area ... Because Knowledge Leads to Better Health!

Please check our website for more information on each of these events as further details become available. <a href="https://www.canpku.org/events">www.canpku.org/events</a>

John, who was diagnosed with mild PKU, started having some problems in school when he was 10, and was later diagnosed with learning difficulties. Much later, says John, he learned that this is a critical age when children being to learn executive function, and where children with PKU are susceptible to problems with this. His son was to take a reduced course load through school, and later through University.

However, when John's son asked to be put on a trial of Kuvan, things changed dramatically for him. John was such a significant responder, he was able to come completely off-diet and reported a lifelong 'fogginess' disappearing. He wanted to complete his University degree but was 8 courses short, says John. Incredibly, he managed to complete those 8 courses with straight A's, double his normal course load.

John feels grateful that his son is a "complete responder" to Kuvan and that they know "his brain works better, including the ability to plan, organize, multi-task and most importantly concentrate in school," because of it. He also says that he appreciates that "no single treatment for PKU will work for everyone," which is why "CanPKU must advocate for every patient and clinician to have access to every tool there is or will be in the PKU toolbox," so that they can all reach their full potential.

\* 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!

#### **BC PKU Event**



Saturday, February 23rd, was BC PKU Day! For a video link to the conference <u>click here!</u>

This exciting event was attended by over 70 people and the line-up of speakers and activities made for a full and exciting day.

The first speaker was Kari Kilmer, a person living with PKU who is also the mother of a child with PKU. Her story of growing up on a ranch in Wyoming and dealing with the daily life on the diet was moving. Dr Sylvia Stockler known by many of us here in BC at the Biochemical Diseases Clinic was the next speaker and gave an informative talk on treatment with Kuvan, it's effects, uses and the definition of what a 'significant' response in those who are tested with it.

Also before lunch, MLA for Kootenay East, Mr Bill



Bennett was awarded a Certificate of Appreciation from CanPKU (see image below). Mr Bennett has been a strong advocate and voice for the PKU community within the BC government. Mr Bennett has been advocating for

person's with PKU for some time now and it is to be hoped with his help, and that of other MLA's who we reach out to, that PKU patients will one day see access to all the treatment options they deserve.

After lunch, Dr Barbara Burton, head of the Chicago Children's Memorial Hospital, spoke at length and detail regarding patient outcomes on Kuvan, upcoming

### **Featured Recipe**

## Jeez-It's\*



The low protein version of Cheez-its, and also similar to Goldfish crackers, this will be a great alternative for any lunch box or snack time! I take out the Cayenne pepper (for the kids).

#### Ingredients:

- 2 tablespoons annatto seed
- 2 tablespoons shortening
- 130 gm baking mix (1 cup)
- 60 gm tapioca starch (1/2 cup)
- 1 teaspoon xanthan gum
- 1 teaspoon sugar
- ½ teaspoon baking soda
- ¼ teaspoon cream of tartar
- ¼ teaspoon salt
- ¼ teaspoon white ground pepper
- ¼ teaspoon mustard powder
- ¼ teaspoon onion powder
- 1/8 teaspoon cayenne pepper (optional)
- 20 gm Molly McButter Cheese Sprinkles (1/4 cup)
- 2 tablespoons butter, cold and cut into ¼ inch cubes
- 60 gm rice milk
- 60 gm heavy cream

#### Directions:

In a small pot, over low heat add

treatments and further on the challenges patients face being both on and off diet.

The President/CEO of the Canadian Organization for Rare Disorders, Mrs. Durhane Wong-Reiger, spoke next about the Rare Disorders network and the importance of advocacy in numbers. Ryan Clarke, President of Advocacy Solutions followed, giving a talk on how best to advocate for PKU patients and their treatment needs with the government and spreading awareness in general. He also spoke at some length regarding the Brain Protection Strategy that CanPKU delivered to the BC Government the day before BC PKU Day.

Last but not least, local dietitian Keiko Ueda challenged our knowledge of all things PKU with a highly interactive game of audience Jeopardy. Who knew that spelling Phenylketonuria could be so tricky (p.s. I hope I got it right)!



Finally, we must mention the wonderful daycare staff - we had 4 babies and many other children in attendance. The team had their hands full but they were very competent and kept the kids entertained all day!

Thanks to all the speakers for presenting as well as vendors (Vitaflow, Cambrooke Foods, Nutricia, Applied Nutrition) who attended with many food samples for us all to try.

Also a big thanks to Local Event Coordinator Sally Broadbent and our National Events Coordinator Jenn Pino. Also, let's not forget our volunteer photographer Amanda Cosburn for many wonderful pictures! If you would like to see more pictures from this event please go to <a href="https://www.pkuamanda.com">www.pkuamanda.com</a>

The BC PKU event was truly a great success and left us all with a wealth of information to digest - **Because Knowledge Leads to Better Health!** 

shortening and annatto seed. Simmer for 10 minutes, strain seed and put mixture in the freezer or fridge until firm (at least 30 minutes). (You can keep the seeds to use again later.)

Meanwhile in a small mixing bowl, combine the rice milk and heavy cream, set aside.

In the bowl of your food processor, combine baking mix, tapioca starch, xanthan gum, sugar, baking soda, cream of tartar, salt, white pepper, mustard, onion powder, cayenne pepper, and Molly McButter. Pulse for 10 seconds to combine.

Add butter chunks and cold orange shortening. Pulse for one second intervals until the mix resembles coarse crumbs.

Add rice milk and heavy cream mixture. Process until a ball of dough forms in the bowl, about 10 seconds.

Turn dough out onto a lightly starched sheet of parchment paper, adding more wheat starch as necessary to keep dough from sticking. Knead dough by gently folding and pressing for 10 to 12 strokes until dough is smooth. Roll dough into a 9 inch square. Fold dough into thirds to form a 9 x 3 rectangle (like a business letter) and divide in half. Wrap in plastic and put in fridge for an hour.

Preheat oven to 300 degrees. Dust a sheet of parchment paper with wheat starch. Roll each half into a 12 inch square. The dough should be about 1/8 to 1/16 inch thick. Using a pastry or pizza

## **Prairies PKU Day is Approaching!**

Prairies PKU Day is coming up soon - Saturday, April 27th, 2013!

Featured speakers include Laurie Bernstein and Dr Robin Casey.

Day care is free and there will be low protein and regular food items available.

Travel scholarships are available for this event so do not hesitate to apply for one should you wish to attend and finances are restrictive.

To register or to find out more information about this event please go to **Prairies PKU Event!** 

## **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.

This year CanPKU is having a national penny drive to raise funds. Collecting pennies from your friends and family members is also a great way for you to spread awareness!

If you have any pennies lying around please consider collecting these to donate to CanPKU. We encourage all members to hold penny drives - at work, school or otherwise to help raise funds for important activities that will benefit all those with PKU.

For more details on how to help please click here!

Your support is greatly appreciated!

wheel, cut dough into 1 inch squares (or use a cookie cutter and shape as desired). Use the blunt end of a wooden skewer or a cut toothpick to poke a hole in the center of each cracker.

If desired sprinkle with a very light coating of salt. Carefully transfer parchment paper to a baking sheet. Bake for 20 minutes.

Mix the crackers around a bit to separate and bake for an additional 15 minutes. Take one out and try it - if it is still a little soft in the middle, bake an additional 5 minutes - it all depends on the thickness of the dough. Let cool on the baking sheet and transfer to wire rack.

Repeat with remaining dough or freeze for later use.

Yield: about 250 one inch crackers

Phe Amount: total 215 mg phe, about .9 mg per cracker

\*taken from Cook For Love by Brenda Winiarski. Please see Cook for Love for more great low protein recipes, and many thanks to Brenda for allowing us to share these in this newsletter!

## Hat's Off To ...

Ontario's Diane McArthur, Assistant Deputy Minister and Executive Officer of the Public Drug Programs, and Deb Matthews, Minister of Health and Long-Term Care.

Both of these people were instrumental in getting Kuvan publicly funded under the Ontario health care system.

## **Update on the BC Advocacy Campaign**

On February 4th, CanPKU representatives John Adams (President) and Nicole Pallone (Vice President) put their families aside and flew to Victoria, BC, for two different meetings with Ministry of Health staff.

BC currently provides the LEAST amount of funding for low protein foods in all of Canada (a \$40/month stipend is available but only to patients who are on social assistance, and only if they figure out how to apply!)

Kuvan is also not funded for PKU patients in BC, although it is purchased by the BC government at full retail price for diagnosing and treating Biopterin Deficiency!

Sadly, both these meetings were unproductive. At the first meeting, regarding Kuvan, we were told in no uncertain terms that the Ministry had completed it's review, decided no, and that only BioMarin could reapply and submit new information. At the same time, they acknowledged that Kuvan helps lower phe levels in some patients, and that low phe levels prevent neurocognitive complications associated with PKU...

The second meeting with Ministry staff regarding funding of low protein foods was also disappointing. Within minutes of sitting down, we were told that we had - once again - been directed to the wrong department and that the people we were meeting with had no jurisdiction over funding of these medically-necessary foods. This is the second time since Pallone started campaigning for food coverage back in July of 2009 that we have been directed to the wrong department - it demonstrates an appalling level of incompetence within the Ministry of Health, and an outrageous waste of both CanPKU and taxpayer funds.

Stay tuned as things develop. We are learning big lessons in BC about how to advocate effectively and although we need to stay focused on one province at a time due to our limited resources, you can bet that at some point CanPKU will be advocating in your province!

After years of advocacy, Ontario recently listed Kuvan (Sapropterin) on the publicly funded drugs list for a subset of patients who meet specific criteria.

Funded under the exceptional access program, this became effective on Feb 28th, 2013, which also coincided with the 6th Annual International Rare Disease Day.

Ontario is now the first province in Canada to publicly fund the drug for PKU patients.

### **Volunteers Required!!**

CanPKU needs your help!

CanPKU is looking for both a volunteer Secretary and a volunteer Administrative Assistant to help support our activities.

The Secretary role will involve attending the teleconference board meetings and taking minutes, typing them up and then distributing for comments and editing. The meetings are quarterly so the expected volunteer time be commitment would approximately 6 hours once every 3 months.

The Admin Assistant is a new role and the exact duties involved are still being developed but the time committment for a volunteer would be in the region of about 5 hours per week on CanPKU related activities.

Without the help of dedicated volunteers there is no question that CanPKU could not undertake all the

We need your voice. Please go to <a href="http://www.canpku.org/email\_premier/">http://www.canpku.org/email\_premier/</a> and sign a letter to the BC Premier about the need for funding of PKU treatments. It literally only takes ONE minutes. Please also ask your friends and family members do the same by emailing this link, posting it on your facebook page and on your twitter account.

Alone our voices are small but together we will be heard!

## Patient Support Project - Starter Kits Round 2!:



We are thrilled to announce that in January 2013 our second round of Starter Kits were distributed to PKU clinics across the country. This round of kits was made simpler and a little more generic, so that a wider variety of patients could be supported.

Now these kits can be handed out to newborns diagnosed with other inborn errors of metabolism, and also to late diagnosed patients or even patients returning to diet.

On behalf of CanPKU, I would like to personally thank the extraordinary volunteers who helped me assemble, package and distribute the kits this time around: Heather Shayna from Manitoba, Tanya Chute-Nagy from Ontario and Tania LeBlanc from New Brunswick, thank you for donating your time to this worthwhile project!

activities and events that it currently does. Your help would be greatly appreciated!

For more information or to volunteer for either of these positions, please contact:

Nicole.Pallone@CanPKU.org

or

newseditor@canpku.org

## Receiving This Newsletter Twice ...?

Did you receive this newsletter twice? That means we have two email addresses for you!

Please email us at the address below and let us know which of your email addresses is the best one to use!

newseditor@canpku.org.

#### **Quick Links...**

**Our Website** 

**Our Previous Newsletters** 

**Our Facebook Page** 

#### **WALKATHON HOSTS NEEDED!**

As mentioned in the last newsletter, 2013 also marks the 1st National CanPKU Walkathon! This will be held on Saturday, June 1st, across the nation. We need hosts for multiple walkathon venues across Canada, to celebrate the close of PKU Awareness Month and to raise funds for CanPKU, to ensure that we can continue offering support and education to families in the years to come.

Our goal this year is to raise \$20,000 at our first National Walkathon, and to do that we need YOU to host a walk in your area! Even if you are the only local PKU family, you will be surprised at how easy it is to organize a walkathon and how quickly small donations from friends and family add up! This is a great way to both raise awareness in your community about PKU and to support CanPKU so that we can continue providing educational opportunities, patient support programs and advocacy efforts on your behalf.

Organizing a walk is super easy: all you need to do is find a location and ask permission, then fill out our form so we know when and where it is being held. City-owned properties sometimes require permits but in smaller communities it often just takes a single phone call!

Please visit <a href="www.canpku.org/walkathon">www.canpku.org/walkathon</a> for more information about how to host a walk or what venues are already lined up!

#### **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 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.

## Dear Reader;

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 <a href="https://www.canpku.org">www.canpku.org</a>.

Sincerely,
Canadian PKU & Allied Disorders Inc.



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