



# CanPKU News

Issue No. 10

Canadian PKU & Allied Disorders Inc. Newsletter

December 2012

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## Dear Reader,

Welcome to the December edition of the CanPKU newsletter!

The final newsletter of 2012, the year ending shortly and a new one about to begin, has made me think back on everything that has happened in the last twelve months. As a family, we had a baby and learned she had PKU, something which we certainly knew very little about (in fact more like nothing) before having her! I became the editor of the newsletter not only to learn more about PKU, but to be helpful and active in the community. It's been an eventful year for our family, and a busy one for CanPKU as well. I can hardly believe it was actually 3 months ago that I sent out the last newsletter!

I imagine that 2013 will be a busy and active year as well. I hope, as I'm sure you all do, that those of us at CanPKU and the community as a whole will be successful in achieving many of the goals we have set to help those with PKU and Allied Disorders.

Please see below for exciting news about upcoming events anticipated for the year ahead as well as our regular articles - the Profile, recipe selection, highlights of the Quebec PKU event (which sounds like it was a wonderful success), and much more besides!

As always, if you have any suggestions or comments regarding the newsletter, I invite you to contact me at your convenience using the reply e-mail option.

Season's Greetings to all!!

Sincerely,

Ruth Appanah

Editor

CanPKU and Allied Disorders Inc. Newsletter



Innovation in Nutrition

### PKU Profile\*

**Amanda Cosburn and  
Amanda Jammeh**



In a twist on our regular profile, this quarter we are highlighting the story of the two Amanda's.

Amanda Cosburn and Amanda Jammeh both reside in Kamloops, BC. Both are 26 years of age and are young adults living with PKU. Amanda

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

As 2012 draws to a close we are already well underway with planning our 2013 Education & PKU Community Events. These events are a wonderful way to connect with other PKU families and to learn more about current treatments and research projects. Although they are not all yet set in stone, these are the dates and locations for the events this year:

Saturday, February 23rd: Vancouver, BC  
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

Stayed tuned for more information in early January about the final dates, locations and speakers lined up for these great events and remember: we try to make travel scholarships available for every event, so don't let financial worries stop you from attending!

May is PKU Awareness month but to make it official in each province requires the help of PKU families like you. Email us today to find out how you can help!

2013 also marks the 1st National CanPKU Walkathon! On Saturday, June 1st, across this great nation we will be hosting a series of walkathons 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. We need volunteer Local Walkathon Hosts in as many towns as possible - we will help you every step of the way and I promise you that it will be a rewarding experience!

If you are interested, please email Nicole Pallone at [nicole.pallone@canpku.org](mailto:nicole.pallone@canpku.org) 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

Cosburn lives with her partner, Cole, and is an LPN. Amanda Jammeh is the mother of two children, both non-PKU, and lives on the north shore.

After having met in clinic a few years back, they have now become good friends after Amanda Cosburn recently moved to Kamloops with her partner.

When asked about what the most difficult aspects of living with PKU are, both Amanda's echo the same sentiment. "The biggest challenge we face is losing weight and the psychological side effects of PKU," reveals Amanda Cosburn. They feel that more support and funding for services for adults with PKU would be invaluable for their psychological needs as well as funding for low protein medical food. Unfortunately, without subsidized help for food, staying on diet is made much harder for them. "With bills, mortgages/rent and daily living, special foods take a back seat," says Amanda Cosburn.

The friendship that these two have developed has been invaluable in helping them through these challenges. As they have both lived through the same challenges, they feel able to support each other through the trials and tribulations of living with the disorder. Amanda Jammeh says "when we first met it amazed me how much we have common and how many similar experiences we have gone through - so many of the same struggles!"

these events as further details become available. [www.canpku.org/events](http://www.canpku.org/events)

## Quebec PKU Event



Saturday, September 22nd 2012 was the Quebec PKU Educational Day and the very first in the Quebec City area, more precisely in the lovely setting of Saint-Augustin-de-Desmaures, on the shore of the St. Lawrence River. We had a great turnout of more than 50 people, almost a quarter of which were PKU adults and teens. Marie-Hélène Bourdages, Dt.P., nutritionist at the CHUQ-CHUL, gave an excellent presentation on Pregnancy for Mothers with Phenylketonuria (PKU) - a topic close to her heart- and also made sure that all her patients and families were well informed on the event and provided us with two extra volunteers from her circle of friends!

We also had the opportunity to hear Virginie Roy-Girard, M.A.Ps, a resident psychologist in the New-Brunswick PKU Case Management Program of the Dr. G. L. Dumont University Hospital Center in Moncton, speak on Living with PKU: From diagnosis to adulthood. She touched on the ways patients and their families deal with PKU at various life stages. This was a rare opportunity for us to explore the psychological aspect of PKU.

Steve and Daniel Dion, two brothers in their thirties both with PKU, testified with much openness and a good measure of humour on their recent experience of returning to diet after 30 years of a diet-free life. They belong to the first generation of PKUers screened at birth who were taken off diet around the age of 6 to 8. Their testimony was very revealing to all present, and they were literally bombarded with questions.

Geneviève Lafrance, nutritionist at the CHUS Fleurimont, in Sherbrooke, closed the day with a presentation on Travelling Tips and Advice that covered just about every aspect of planning a PKU-friendly trip or outing, from what to pack, cruising through Customs, air traveling, all

Together both Amanda's are working to improve their health by doing things such as taking regular walks together, getting together to bake low protein foods, and supporting each other in their daily lives. The results have been wonderful. With a return to diet, Amanda Jammeh's Phe levels have gone from over 18 to most recently just under 9, while Amanda Cosburn's levels have gone from 8 to 2!

Another thing the two Amandas have done together is to create a blog site documenting their life and challenges, where they are also keeping a daily record of their goals and Phe levels. To read more on their story - click here:

### [The Two Amanda's](#)

\* 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!

## **RECIPE CORNER**

### **Low Protein Gingerbread Men Cookies\***

inclusive vacations, Disney World and eating out in restaurants. Attendees then shared their own experiences which, ultimately, demonstrated that you can certainly take PKU on the road!

The kids in daycare had a ball with Mireille doing crafts, playing games and even creating a play. We can only regret that they shied away from performing it for the rest of us. It was yet another fantastic day that truly echoed the CanPKU motto: Because Knowledge Leads to Better Health.

## **Canadian Orphan Drug Network**

The Canadian Government, on October 13, 2012, released a statement relevant to all people affected by rare disorders. The government announced two initiatives of import - the creation of a new approach for the authorization of "orphan drugs" and also the Canadian launch of Orphanet - a comprehensive on-line database of information and services for rare diseases.

The Honourable Leona Aglukkaq, Minister of Health, was "pleased to announce that [the] Government will introduce a new approach that will better support the development and authorization of drugs for rare diseases and launch a new Web portal to assist patients in finding the information and services they need." This new approach, with a regulatory framework for use and guidelines on orphan drugs, has been created to allow patients suffering from rare disorders to better access the services and medications they require.

Orphanet, meanwhile, is an international on-line reference portal with information on the diagnosis, care, and treatment of patients with rare diseases. With funding from the Canadian Institutes of Health Research (CIHR), this is now available in Canada.

CanPKU and, indeed, many in the PKU community have for years been advocating for the need for a national strategy on treatment of rare disorders. It is hoped that this new framework and launch of Orphanet will ensure increased support for the rare disease community in Canada and allow those with PKU access and funding to every treatment option they require.



#### Ingredients:

1  $\frac{3}{4}$  Cups Baking Mix (Cambrooke foods)

1 Cup Wheat Starch

2 tsp Baking Soda

2 tsp Cinnamon

1  $\frac{1}{2}$  tsp Ginger

1  $\frac{1}{2}$  tsp Cloves

$\frac{1}{2}$  tsp Salt

1 Cup Butter (162 gr.)

1 Cup Brown Sugar (lightly packed)

$\frac{1}{4}$  Cup Molasses

#### Directions:

1) Combine 1st 7 ingredients. Mix Well.

2) Cream Butter, brown sugar and molasses until it is light and fluffy (Using hand held mixer).

3) Add dry ingredients gradually to the creamed mixture, mix well. (Using hand held mixer) Chill dough for easy rolling.

4) Roll dough out (1/4 inch thick)

For the complete press statement please click here: [\(English\)](#)  
or [\(French\)](#)

To visit the Orphanet CA website please click here:  
[\(English\)](#) or [\(French\)](#)

For CanPKU's statement on Health Canada's initiatives please click  
here:  
[CanPKU Orphan Disease Framework statement](#)

### **BC Media Campaign Kicked off in October**

As you all know, we have work to do in each and every province when it comes to improving the funding of PKU treatment and clinical services. For almost two years now we have been actively engaged with both the BC and Ontario Ministries of Health as they review whether or not to fund Kuvan for PKU patients. We have also been involved with the Quebec and Saskatchewan Ministries. We are happy to report that Ontario is still actively engaged in negotiations with BioMarin, the manufacturer of Kuvan. Unfortunately, the same cannot be said in BC where the Ministry of Health completed their review this summer and chose not to fund this innovative treatment for PKU. As such, our advocacy efforts have focused primarily on BC in regards to funding both Kuvan and low protein foods (in BC, only patients are social assistance are eligible to apply for a monthly stipend of only \$40 per month).

In October, we launched our media campaign in BC which has been a great success. We have had several articles published in BC newspapers, some subsequent letters to the editors of those papers and a few radio interviews as well. To view the media coverage please click here: [Media Coverage BC](#)

Extra special thanks to the families of BC PKU patients Svenga Fortstrom, Alayna Lemky, Rosie Pallone and Connor Laing who have all made efforts to meet with their local MLA. When an MLA or a group of MLA's can put pressure on the Ministry of Health behind the scenes it is a huge help, and it is the job of an MLA (or MPP in Ontario) to listen to you as constituents and stand up for your needs. We are still looking for more PKU families to speak out and meet with their MLA's to keep the momentum going, especially in areas like Victoria, BC, where we had media interested but no local family!

onto lightly floured (wheat starch) board.

5) Cut with cookie cutter and place on greased cookie sheet. Or use parchment paper.

6) Bake at 325-350 degrees for 8-9 mins or until set. Cookies may appear uncooked when you take them out of the oven, but they will set quickly. A perfectly done cookie will be slightly soft when it is cool.

Nutritional Information:

Makes 26 cookies @ 30 g each

Per Recipe: 6.30 exchanges (94 mg Phe)

Per 30g Cookie: 0.25 exchanges (<4 mg Phe)

Wheat Starch (1 cup) = 0.50 exch., Baking Mix (1 3/4 cup) = 0.18 exch., Butter (162 g) = 4.32 exch., Cinnamon (2tsp) = 0.40 exch., Cloves (1 1/2 tsp) = 0.45 exch., Ginger (1 1/2 tsp) = 0.45 exch., In Total: 6.30 exch/recipe

\*note that this recipe was kindly provided by Brienna Young

We also launched our online advocacy tool that makes it super easy and quick for anyone to send an email of support and within a month or so we had over 500 sent in to Ministry of Health officials and Premiers across the country. This tool is meant to be used by everyone including patients, families, friends and colleagues as regularly as once a week. You can edit it as you see fit so that it is specific to the needs of your province - see how quick and easy it is by clicking here:

[Email your Premier and Health Minister!](#)

On Monday, December 3rd, BC resident and CanPKU Vice President Nicole Pallone was able to meet with the BC Minister of Health. This meeting was arranged through the insistence of Pallone's local Member of the Legislative Assembly (MLA) Mr. Bill Bennett and highlights the critical need of provincial residents to become involved in any advocacy campaign we undertake in their province, including being involved with their own provincial MLA/MPP/MHA/MHA. At this meeting, Minister MacDiarmid said that the Ministry would re-evaluate the possible funding of low protein foods and that they would also review their decision on Kuvan when new information became available. Although we are always hopeful for a positive decision, we are encouraged by their willingness to continue reviewing these issues, and we look forward to future meetings with BC Ministry of Health staff that have been scheduled.

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 - and we will need your help!

We need your voice. Visit our website at [www.canpku.org/advocacy](http://www.canpku.org/advocacy).

## Hat's Off To...

**The Rose Award Winner**

**Brenda Winiarski**



## **Biomarin Announces Decision to Start Phase 3 Program for PEG-PAL in 2Q 2013**

On Sept. 26, 2012, in GLOBE NEWSWIRE, Biomarin released the results of its Phase 2 study on the drug PEG-PAL, used in the treatment of patients with PKU.

"This Phase 2 study has shown that PEG-PAL appears to control

The Rose Award is an award given by Applied Nutrition, based on nominees from the public or community to recognize an individual who makes a consistent and outstanding effort in giving to the PKU/MSUD community.

Brenda Winiarski, PKU chef extraordinaire and creator of the culinary website and recipe book Cook For Love, is this year's winner. Cook For Love is a non-profit website dedicated to improving the health of people in the PKU community through cooking and education and to dispel the myth that low protein cooking is not equal to or as appealing as that of its regular counterpart. Anyone who has tasted a Cook For Love recipe can attest to the fact low protein foods can be truly delicious. We are ever so grateful that Ms. Winiarski, even as a busy Mom, chooses to share her love of cooking so that we can all Cook For Love.

Please be sure to check out her website, sign up at no charge for access to her recipes and check out the Cook For Love Facebook page!

For more information on this award and this year's winner please click here:

[Rose Award Winner](#)

To visit Cook For Love please click here: [Cook For Love](#)

To visit the Facebook page for Cook For Love, please click here: [Facebook Cook for Love](#)

Phe levels independent of the Phe-restricted diet and can produce a sustained response," stated Dr. Nicola Longo, Professor of Pediatrics and Adjunct Professor of Pathology at the University of Utah and Clinical Investigator in the PEG-PAL Phase 2 trial.

According to Biomarin, of the 25 patients who have been treated with PEG-PAL for at least one year, blood Phe measures were on average 68% lower than pre-treatment baseline levels. Furthermore, patients had blood Phe measurements below 600 umol/L - well below the the NIH guideline of 900 umol/L. In addition, all patients tolerated the drug well except for exhibiting hypersensitivity-type reactions consisting of injection-site reaction at the site of administration. This hypersensitivity, states the company, can be ameliorated with a personalized dosing regime started at minimal amounts before escalating to a maintenance dose of the drug.

Based on the outcome of these results, Biomarin also announced its intention to start Phase 3 trials of the new PKU treatment drug PEG-PAL starting in the 2nd quarter of 2013, following an end of phase 2 meeting with the FDA in the first quarter of 2013.

To read the complete press statement please click here: [Biomarin Press Release](#)

## Become a Member!

The more members we have the greater our voice is when advocating for better treatment coverage and care.

To become a member go to:

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

**Please note that the annual membership fee for professionals has been reduced to \$25 per year.**

Approved at the AGM in July 2011 and subsequently approved by Industry Canada in July 2012, our Membership Categories have been updated as follows. Please note that all members in good standing who have voting rights will be re-classified as General Members so as to retain their voting rights. Please

## Special Thanks To:



Many of you will have recently received an advent calendar in the mail. These were graciously donated to CanPKU by Nutricia Metabolic Nutrition.

110 calendars in total were sent out this year all across the country.

Didn't get one? Become a member and you won't miss out on future goodies! (see our becoming a member section to find out about the benefits of joining)

Thanks to Nutricia for this generous donation!

## 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](mailto:newseditor@canpku.org).

## Quick Links...

[Our Website](#)

[Our Previous Newsletters](#)

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.

**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!**

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.

Becoming a member includes the following benefits to you:

- Direct contact from CanPKU regarding newsletters and event invitations;
- Direct access to new information regarding treatments, research and Provincial/Territorial advocacy campaigns;
- Support from other individuals and families who understand;
- Reduced registration fees for all CanPKU events;
- Priority access to travel bursaries for CanPKU events, when available;
- Opportunities to volunteer and make a difference;

[Our Facebook Page](#)

- Tips on advocacy and creating awareness in your community; and
- Voting rights at Annual General Meetings and Special Meetings (General Members Only).

[Join Our Mailing List!](#)

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 [www.canpku.org](http://www.canpku.org).

Sincerely,  
Canadian PKU & Allied Disorders Inc.



Because Knowledge Leads to Better Health

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