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CanPKU News

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In This Issue

PKU Profile

Gabrielle Laplace

Gabrielle was born in Montreal, QC on February 26, 1985 and was diagnosed with PKU when she was 3 weeks old. She was the first in her family diagnosed with the disorder, although an uncle in the 1930's was diagnosed as an 'idiot' and may have had PKU as well. Gabrielle's experience with school ranged from being a straight 'A' student to having real trouble with certain tasks and she was diagnosed with a learning disability during her elementary years. It has been difficult at times to cope with the strict diet and with trying to explain quickly and simply what PKU is, but because of her experience Gabrielle has grown into a young woman who is very tolerant of people who have conditions that set them apart. Gabrielle has successfully completed her post-secondary education and is currently working in the equestrian industry, which she loves. When asked about the PKU diagnosis, her mother Helene said they took it in stride. "We had the good fortune of

2011 Remaining Events:

July 8th & 9th: Ontario PKU Event & Polo4Kids, Alliston, ON
July 10th: Annual General Meeting, Alliston & Teleconference
August 27th: Please note that NL PKU Day has been rescheduled (originally set for June 4th)
September 18th: BC PKU Day, Vancouver, BC
October 15th: Quebec PKU Day, Montreal, PQ

Article about CanPKU Event in Red Deer, AB

by Laura Tester of The Red Deer Advocate
Published: June 13, 2011 9:34 AM

Alberta families impacted by rare disease press government for more financial help

The Alberta government is being pressed to put more money back into treatment of children who are suffering from a rare inherited metabolic disorder.

Families do receive some free coverage for foods designed to

being told by a very experienced and tactful nurse. The support from the hospital staff in the weeks, months, and years that followed was precious." She points out that the diagnosis of her daughter was 'pre-web' so all of their knowledge about PKU came from the hospital staff; much different from many of todays' newly diagnosed parents who jump online and read about all the negative effects. As a parent, Helene got tired of being the 'food police' but watched her daughter develop a maturity about her diet as she grew up until the diet was fully in Gabrielle's hands. "The diet itself is both the best and the worst part of having PKU" says Helene. The diet is difficult to follow and hard to explain, but it is wonderful that it can successfully minimize the negative effects of PKU. In the last decade, there has been more focus on the links between PKU and neuro-cognitive difficulties, even in those who successfully manage their levels well with diet. "In hindsight, Gabrielle's PKU probably contributed to the times in her schooling when she did struggle, but until recently there was no evidence to support the theory." Regardless of some rough spots, Gabrielle pushed herself with tough programs. She took a break during college and moved to Alberta for a year to work at an equestrian center, and then returned to Quebec to finish her education. At 26 Gabrielle is living with her boyfriend and while she is not working in the field she went to school for - as happens to so many of us - her Mom thinks that is OK. "I am happy that she is doing what she loves - it makes her happy and keeps her healthy."

Gabrielle Laplace's mother, Helene Dandurand, is CanPKU's Director of Francophone Relations.

keep their children, who have PKU, from sustaining brain damage. But that list of foods has been reduced. Plus, there is promising drug therapy that needs to be considered for coverage as well, say families.

On Saturday, about 60 people from across Alberta attended Canadian PKU and Allied Disorders group's Prairie PKU Fun and Education Day at Gaetz Memorial United Church on Ross Street. About one in 12,000 Canadians are born with PKU, short for phenylketonuria - a disorder that must be treated within the first few weeks of life or else the individual can suffer mental retardation and other neurological problems. A standard blood test is done when a child is born.

Sufferers are missing the enzyme phenylalanine (PHE) that's found in protein and if there's a buildup of PHE, it becomes toxic and causes permanent brain damage. Most PKU children and adults follow a special diet that involves a strict control of natural protein intake, eating special low-protein food and drinking a synthetic PHE-free formula. The formula and food are expensive.

John Adams, president of the Canadian PKU and Allied Disorders group, said he'd like to see the Alberta government be as generous as the government of Ontario towards paying for formulas and specialty foods. "Unfortunately, your province has recently cut back on the list of products that are available for PKU families," he said.

He'd also like to see all provinces cover new drug therapies for those with PKU. The government of Quebec recently announced it would pay for a category of PKU patients so they can take drug therapy.

Jerry Vockley, chief of medical genetics at the Children's Hospital in Pittsburgh, Pa. where his clinic treats more than 200 PKU patients, said the new drug therapy called sapropterin is offering hope. Taken in pill form, it's been approved in the United States, but is "struggling to gain acceptance here in the Canadian medical system."

"We've gone from nothing to offer these patients, to having half of them reduce their (PHE) levels and having 10 per cent of them become completely normal," Vockley said. "Increasing your ability to eat just a little bit of (PHE) makes a huge difference in your diet."

Event organizer Sandra Harland said more than 200 families in Alberta have children with PKU. Her 12-year-old daughter is good with her diet, but on the occasion she gets too much PHE, she will have mood swings and increased hyperactivity.
"We are saying to the government that if you will pay for this

Travel Scholarship Available

Apply for a CanPKU travel scholarship and join us in Alliston, Ontario on July 8-10 for our CanPKU Polo-4-Kids event and our AGM! Applicants must be at least 19 years of age and be residents of Canada. Preference will be given to CanPKU members and those with or parents of those with PKU. For more information about the event, click here. Or, apply now!

Hat's Off To...

Local Event Organizer (LEC) **SANDRA HARLAND** for all her hard work in organizing the event on June 11th in Red Deer, AB.

The event was a great success with 22 different PKU families in attendance and a total of 77 people. The speakers were excellent and the children were entertained by a magician in the daycare. There was ample food (for both PKU diets and non PKU) and the atmosphere was relaxed and friendly. Everyone in attendance was able to ask questions of the experts and learn about the products of our sponsors. It was a great day all around - thanks Sandra!

We rely heavily on our LEC's to help with all aspects of planning and running our events. If you want an event in your area please contact Janine Anderton and let her know you want to be an LEC!

long term, it's going to be less expensive," Harland said. "If the children go off the diet, there's going to have to be long-term care that the (government) will have to pay for."

Red Deer's Amy and Ross Christensen, both carriers of the PKU gene, have a five-year-old daughter with the disorder. Their younger daughter doesn't have it. Amy is now pregnant with her third child, who has a 25 per cent chance of having the disorder. They order their special formula and flours through an Edmonton clinic. Any extra food they want comes from Toronto Sick Kid's Hospital, or a company in the United States. A loaf of bread can cost $17.

Amy feels that Alberta has some of the best health coverage in Canada for PKU supplies. "Having said that, things have changed where we used to have access to a lot more products," she said. Ross said there's less variety as a result. For instance, fake chicken nuggets that gave children some "normalcy" are no longer covered.

The couple is excited about the latest medical research, though. "We always hope for a better treatment than just the diet," Amy said.

Sheila Jones - 60th Anniversary of Successful Treatment for PKU

Sheila Jones was born on October 1, 1949 and was referred by her family doctor to a special clinic at Birmingham Children's Hospital, England where she was seen by Dr. John W. Gerard, a pediatrician, on March 13, 1951. (Dr. Gerard is 94 years old, living in Saskatoon, SK.) She was 17 months old and "could sit only and rocked continuously," according to his case notes. A urine sample was tested by Dr. Horst Bickel, an MD from Germany doing post-graduate studies in biochemistry, and she was diagnosed with PKU.

The **Applied Nutrition ROSE Award** (Recognition of Service Excellence) recognizes individuals who consistently demonstrate outstanding service and selfless work to help improve the health and welfare of those with PKU or MSUD.

The winner of the ROSE Award will be honored with a $1000 donation in their name to the PKU or MSUD organization, camp or clinic of their choice. In addition, the award will be presented at an event held for their local clinic or organization, sponsored in full by Applied Nutrition.

Appropriate nominees include leaders, volunteers and members of local, regional, state, national advocacy or support organizations, parents and adults with PKU or MSUD who work to improve the health and welfare of those living with these disorders and metabolic professional.

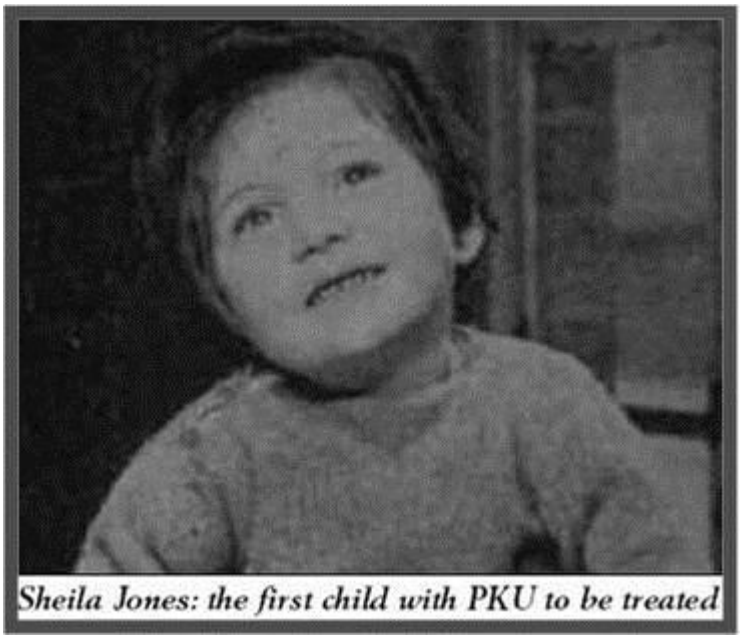
To submit a nomination or learn more about the ROSE award, please go to:
www.medicalfood.com/ROSE

Recipe Corner

Vegetable Curry

This delicious, quick and easy dinner can be enjoyed by the whole family!

Makes 4 adult servings with 5.56 exchanges or 84mg of Phe each. Total recipe: 22.21 exchanges or 333mg Phe. Preparation time: 10 min. Cooking time: 15 min.



Sheila Jones: the first child with PKU to be treated

Her mother, Mary brought Sheila to the clinic almost every day for six months, with or without an appointment, insisting they do more than just diagnose her. Sheila was admitted to hospital for two weeks in September for observation and admitted again in November when she was "put on a phenylalanine-free diet". On December 18, the diet was changed to a low-Phe diet due to weight loss. She remained on the diet in hospital until May, 1952 when she was sent home to be seen at clinic once a week where her mother received the prepared formula. There was a notable improvement on diet.

The low-phe diet, using an amino acid formula made by filtering Phe out of a milk product called casein, was the idea principally of Louis Woolf, a biochemist working at the Children's Hospital in London, England, who taught the professionals in Birmingham how to make the formula. Dr. Woolf is 92 and living in Vancouver, BC.

In October 1952, without the mother's knowledge, the prepared formula was changed to include 5 grams (5,000 milligrams) of phe - about 150% more than a child of her age without PKU would receive for a regular diet. "By nightfall, she was crying and banging her head as she used to do," the case notes report. "On the following day she cried continually and her eyes looked heavy. She could no longer stand while holding onto a chair and her crawling was weak. Her mother felt there must have been a mistake with the preparation of the diet and discontinued it. On the following Wednesday...Mrs Jones came for the first time without Sheila and in tears, because she said something had put Sheila back to where she had been when we first saw her, and that she was unable to stand and was quite lifeless..." Sheila was placed on the proper diet and in about a week "seemed back to normal."

Ingredients:

70g Onion

10g Garlic

120g Bell Pepper

120g Celery

140g Green Bean

140g Carrot

30g Apple, peeled & cored

2 tbsp Olive Oil

2 packets G. Washington Golden Broth

24 oz. hot water

3 tsp Curry Powder

Salt & Pepper, to taste

1 tbsp Cornstarch

2 tbsp cold water

Instructions:

Dice first 7 items. Heat olive oil over med-high heat in fry pan. Cook onions and garlic until golden, about 2 min.
Stir frequently to prevent burning! Add green beans and carrots and cook until softened, about 5 minutes. Add Celery and Peppers and cook another 2 minutes. Mix broth packet in 12oz of water and add to fry pan. Add apple, curry, salt and pepper. Reduce heat and simmer to desired tenderness, about 5 minutes. Mix cornstarch with 2 tbsp cold water and add to fry pan. Stir well until thickened. Spoon over rice or pasta (or low-protein rice/pasta) and serve!

Sheila was hospitalized, for the third time, for seven weeks in November and December 1952. This period in the Birmingham Children's Hospital during November and December 1952 is what is shown in the 8 mm silent film. Watch it here. Sheila lived with her mother until Mary suffered a mental breakdown and was institutionalized when Sheila was 10 - she spent the rest of her life off diet. She died at the age of 49, in 1999, of a liver ailment reportedly unrelated to PKU.

Sheila and her mother Mary helped change PKU from untreatable to treatable - and for that we are all thankful.

Volunteers Needed!

Are you good at letter writing? Comfortable with public-speaking? Good with math and like playing with numbers? Do you have experience in marketing, or enjoy thinking outside the box? These are just a few of the skills that we could put to good use!

There are lots of different ways you could contribute, even if you only have a few hours to give. We have one-off projects, jobs that require just a couple hours a week and some larger jobs that are integral to a smooth-working organization.

If you are able to contribute some of your valuable time to us you will experience the great satisfaction of making a difference. Interested? Email Nicole.Pallone@canpku.org for more details! We'll ask you some questions about your skills and match you up with a task that is right up your alley!

Advocacy 101

Advocacy is verbal support or argument for the purpose of instigating action. It is not activism; it is grounded in logical, rational thought. There are differences between awareness, education, and advocacy - only advocacy is about tangible change.

In each newsletter we will provide simple tasks that everyone can do to further the cause, and tips on how to be an effective advocate.

Your task this quarter: write a letter to your local MLA. Try to get them involved. For added punch, write letters to your Minister of Health, your premier, or the opposition health critic in your province. The more these people hear about PKU, the more likely they are to listen!

Tips to make it effective include making it personal and keeping it simple. Tell your PKU story: how it has affected you and your loved ones as well as what needs to be done to make life better for you and yours. Explain a little bit about what PKU is, but keep it simple. Make sure your letter is specific to what needs

Visit Our Sponsors

ASK US!

Do you have a PKU related question that you are having trouble finding the answer to? Send an email to

nicole.pallone@canpku.org with "Ask Us" as the subject: we'll do our best to find the answer!

Join Our Mailing List!

to be done in your province. (For information on this, email us or check out the 'resources' section on our website for a summary of what is covered in each province.)

As with any of your advocating efforts, we're here to help. Feel free to call or email us for assistance in this regard.

Please note: CanPKU fully supports each of its members being personal advocates; however, any communications that show affiliation with CanPKU including but not limited to the use of the CanPKU logo, CanPKU letterhead or your title as a CanPKU member means that the communication must be pre- approved by the CanPKU President or Vice-President.

Become a Member!

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

NEW! As of June 1st all new members will receive one free issue of National PKU News, courtesy of Virginia Schuett!

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;
- Tips on advocacy and creating awareness in your community; and
- Voting rights at Annual General Meetings and

Special Meetings.
To become a member go to
http://www.canpku.org/become-a-member

Join the Dialogue!

CanPKU is actively involved in advocating to the Provincial and Federal governments about PKU coverage and care. We have met with many officials in various provinces and we have made submissions to the Common Drug Review as well as the provincial governments that allow us to do so for the coverage of Kuvan.

Both the President & CEO John Adams and Vice President Nicole Pallone have attended workshops on how to make patient submissions more effected. The workshop below is open to patients and caregivers as well as patient advocacy groups.

John Adams, our CanPKU President, is one of two speakers from patient groups who will share their experiences in providing input to CADTH and provide tips to help others can prepare their evidence submission. It seems the CanPKU submission got noticed.

Patient Involvement in Drug Coverage Reviews

Join us for a discussion on how patient groups can meaningfully contribute to the drug coverage review process. **Hear** from Canadian organizations and publicly funded drug plans that have recently implemented a patient input process and from patient groups who have provided evidence to date. **Share with us** what would help your group file submissions that provide maximum value to the review process.

What: Interactive event for patient groups
When: Wednesday 22 June 2011
Time: 11:00am to 4:30pm, lunch provided
Where: Hilton Toronto, 145 Richmond Street West
How: Complete the attached registration form and return to events@CADTH.ca
Cost: There is no fee for patient groups and the public health sector. Private sector representatives are welcome to observe and a $200 registration fee will apply.