CanPKU News

Issue No. 7

Canadian PKU & Allied Disorders Inc. Newsletter **March 2012**

In This Issue

Dear Reader,

Spring is in the air! We've recently announced our line up of education and networking events for 2012 and hope to see you all there. We have ambitious plans yet again this year and hope more volunteers will come forward - especially those that are fluent in both French and English!

Nous travaillons fort en ce moment pour franciser nos communications. La version française de ce numéro de notre bulletin sera très bientôt diffusée sur notre site Web. Pour la lire, cliquez ici. Nous prévoyons publier le prochain numéro en français et en anglais simultanément.

As always, we welcome questions and comments about PKU and this newsletter so don't hesitate to email us if there is something you are curious about... Because Knowledge Leads to Better Health!

Sincerely,
Nicole Pallone
Vice President & Director

2012 Event Series

CanPKU Education & Networking Dates Announced!

Please join us at the education and networking event in your region to hear fabulous speakers, meet incredible people, try new low-phe foods, meet vendors of metabolic products and more!

As much as possible we will be offering Travel Scholarships to assist those families who can't afford to travel to our events. For more information on the travel scholarship eligibility criteria please **click here**.

The 2012 Event Dates are:

- **Prairies PKU Day: Sunday May 6th, Winnipeg, MB**
- **BC PKU Day: Sunday June 10th, Vancouver, BC**
- **Atlantic PKU Day: Saturday June 16th, Moncton, NB**

RECIPE CORNER

Chocolate Cake

This Easter make a delicious low-protein chocolate cake that the whole family will enjoy! This cake is light and moist and DELICIOUS.

Ingredients

6 tbsp oil
2 tbsp cocoa
1.5 cups of Loprofin flour mix
1 cup sugar
1 tsp baking powder
1 cup of cold water or coffee
1 tsp vanilla extract
1 tbsp vinegar
1 tsp salt

Method

Pre-heat oven to 175 degrees C

Sift the Loprofin flour into a bowl. Add the remaining dry ingredients and whisk together (the whisk is important as it helps to keep the air in the mixture).

Add all liquid ingredients and mix on high with an electric mixer until the batter is nice and smooth.

Pour the mixture into a cake tin or cupcake cases.

Bake at 175 degrees C for 35 - 40 minutes for a cake, or 20 - 30 minutes for cupcakes. The cake is done when a skewer comes out clean. Decorate as desired and don't forget... you can have some fun with it!

This recipe freezes well.

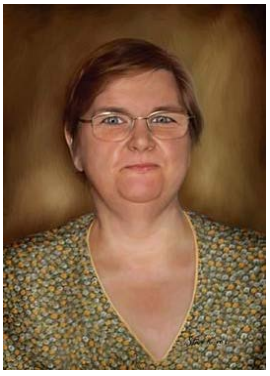
PKU Profile

Hayden Knox

- **Ontario PKU Day: Saturday July 14th, Alliston, ON**
- **Polo-4-Kids: Saturday July 14th, Alliston, ON**
- **Quebec PKU Day: Saturday September 22, Saint-Augustin-de-Desmaures, QC**

Please check our website for more information on each of these events as details become available. www.canpku.org/events

Tribute to Dr. Lambert (1952-2012)



We learned with sadness that Dr. Marie Lambert passed away on Monday February 20, 2012 at the age of 59 years. An active member of the Canadian genetics community for over 30 years, she will be justly remembered for her integrity, determination and vision, and for her unequalled legacies to Genetics training and to the establishment of Medical Genetics as a specialty in Québec.

Following Pediatrics residency training at Sainte-Justine Hospital, which included Genetics electives with Louis Dallaire and Serge Melançon, Marie completed her postdoctoral research training in molecular genetics with Roderick McInnes at the Hospital for Sick Children. She returned to the Université de Montréal and Sainte-Justine in 1986. There she built and maintained an active practice in pediatric medical genetics focused on biochemical genetics, hyperlipidemias and cardiovascular health. She directed the biochemical genetics diagnostic laboratory of CHU Sainte-Justine from 1990 until her death. In recent years, Marie devoted increasing time to the highly successful interuniversity research groups centered on intervention and treatment in child and adolescent cardiovascular health and on the course of insulin resistance, of which she was a founder and leading figure. At the Université de Montréal, Marie was the principal force in creating the Royal College residency program in Medical Genetics (1992) and the Masters of Science program in Genetic Counseling (2003). She played a leading role in the definition and recognition of Medical Genetics as a specialty in Québec, and served as the first president of the Association des médecins généticiens du Québec from 1997 to 2002.

Many with PKU and other IEM disorders treated at Sainte-Justine Hospital, in Montreal, have had the privilege of knowing her and she has seen at least a full generation of them bloom into adulthood. She will be sorely missed by many patients and their families, and remembered as a very dedicated and determined physician who spared no efforts to save and protect them from the ill effects of the disease that threatened them.

We send our heartfelt condolences to Dr. Lambert's family, friends, colleagues and all who knew her.

Advocacy 101

Advocacy is verbal support or argument for the purpose

Almost five year old Hayden Knox of Kamloops BC is a typical bundle of energy boy - typical except for his PKU! Diagnosed at 9 days old, his Mom Brienna Young and Dad Terry Knox were devastated when they heard the news. Young recalls the first meeting at the doctor's office when they were told that PKU could lead to brain damage. "At those words my entire body went numb," says Young. "Even with Hayden fast asleep in his car seat beside me it felt as if the doctor had just snatched away our perfect little baby." Young recalls feeling an immense mixture of emotions including shock, pain, anguish as well as love and protectiveness. She knew immediately that she would do everything possible to keep him healthy.

Hayden's diagnosis has had a profound effect on almost all aspects of Young's life. As a single parent and full time nursing student, Young has often felt like she is the only one who can properly manage his diet and keep him developing to his

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: meet with your locally elected provincial representative. Building relationships with decision-makers is a central part of advocacy and with health issues like PKU that typically means approaching your Member of the Legislative Assembly (BC, AB, SK, MB, NS, NB and PE), Member of Provincial Parliament (ON), Member of the National Assembly (QC) or Member of the House of Assembly (NL).

The first step is to telephone your provincial representative's constituency office (this information can be found online) and tell them who you are, that you live in the riding and that you would like to set up an appointment. Be sure to tell them that you'd like to talk about PKU and ask to meet for 30 minutes. Remember, it's that person's job to meet with you, so don't take 'no' for an answer.

Just before the meeting date, contact the constituency office to confirm the date and time, and let them know who will be attending. Try to go to these meetings with at least one other person.

On the day of the appointment, arrive early. When greeted by your provincial representative, try to get right down to business by telling that person who you are, who you represent (i.e. your child), why you are there and what you need them to do for you. That will set the stage for a very focused and efficient meeting.

Your meeting should then be divided into three parts going forward. The first part will be delivering your key

highest potential. That being said, Young has a very supportive family and she and Hayden live on property adjacent to her parents' farm. Her parents help her out a lot and Hayden's daycare has done amazingly well with his diet.

For Hayden's mom, the hardest part about his PKU is traveling. Even staying overnight somewhere involves packing up formula, a scale, containers for the prepared formula, low-phe snacks and dinner items, measuring spoons, his feeding record, calculator, a cooler and the low protein food list. "It is something that gets easier with time. After 4 years of traveling to Vancouver I think I have it down to an art!"

While traveling to BC Children's for Hayden's checkups twice a year is difficult financially on a student's budget, Young credits his diagnosis with motivating her to go back to school and get her nursing degree. "There are some good things about PKU," says Young. "I feel like you have a special bond with everyone in the PKU community. I don't have to worry about whether or not my child is getting enough vitamins or minerals, and I have access to a team of highly specialized health care workers who are continually assessing your child. The care we have received from BC Children's Hospital is fantastic."

Hayden is, by all accounts, a dynamic little boy. "Everyone who has spent any time with Hayden mentions what a

messages and your one 'ask'. Key messages are the two or three critical things your elected representative needs to know about PKU. Their purpose is to educate the listener. Your one 'ask' is then what you need your provincial representative to do as a result of what you've told them.

The next part is your personal story. Whether you are a patient, caregiver, parent or grandparent, you have a personal story to tell about how PKU has impacted your life. Tell it - with passion and conviction.

Lastly, it is important that you leave time for you provincial representative to talk. Effective advocacy is about presenting, listening and responding. As well, don't be afraid to ask questions or seek clarification, and be sure to establish clear follow-ups with timelines.

Make sure to write a thank you letter or e-mail, and provide any further information about PKU that may have been requested right away. Follow up regularly to track the progress of the 'ask' that you presented.

As with any of your advocacy efforts, we're here to help. Feel free to call or email us for assistance in this regard and please inform us of meetings that you schedule and results of those meetings. It is very helpful (and difficult) for us to have an overview of what advocacy efforts are happening in all the corners of this country!

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.

character he is," says Young. So far, Hayden has done a good job of learning what foods he can and cannot eat and getting him to stick to his diet hasn't been a huge challenge. Hayden goes to daycare 5 days a week and loves to play on his grandparent's farm. He rollerblades, rides his bike, feeds his 12 chickens and collects their eggs. He also has 3 dogs, 3 horses, 2 fish and a cat!

When Brienna Young graduated from high school her goal was to be a waitress in a fancy restaurant. "Knowing that I would do anything for Hayden showed me that I could do anything I wanted to for a career," states Young. Now she is an almost-graduated nurse and on the Board of Directors for Canadian PKU and Allied Disorders Inc. She has been inspired by Hayden to strive for the things that will make her happy, and hopes that he too will realize one day that the possibilities are limitless. At five years old, Hayden thinks being a garbage truck driver would be the best job ever - and that's just fine with his Mom if that's what makes him happy.

PKU Starter Kits

New CanPKU Initiative a Huge Hit

Just before Christmas, CanPKU President Nicole Pallone added to the Canada Post workload by sending out 60 PKU Starter Kits to 20 different clinics across Canada.

These kits contain loads of information and all the necessary tools for successful PKU diet management including The PKU

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.

The History of PKU Formulas

by Tony Parington, with Josh Prizer and John Adams

The Author:

Tony Partington has had over 30 years experience in

Food List, scales, calculators, a diet journal and more.

Nicole Pallone's spare bedroom, filled to the brim during starter kit assembly!

"The purpose of these kits," says Pallone, "is to ease some of the stress that accompanies a PKU diagnosis by providing the essential information and tools needed by families new to the PKU diet. Although this project was aimed at newborns, we learned that more kits could be used for late-diagnosed patients in care facilities and older PKU patients that move here from other countries."

While Pallone hopes to expand the Starter Kit project to include these other patient, the reality is that the project itself is in jeopardy. "Finding a way to make this project financially sustainable is a challenge," says Pallone. "We have limited resources and unless we can secure more sponsorship or donations specifically for this initiative, we may not be able to go ahead with the next round of Starter Kit assembly, scheduled for Fall 2012.

The contents of the 2012 PKU Starter Kit

developing disease specific nutritional formulas and was involved from the 1960s in product development for inborn errors of metabolism. Most of this time was spent in SHS/Nutricia and, more recently Tony helped set up Vitaflo International Ltd. Vitaflo International has become a significant developer and innovator in medical nutrition.

PKU Formula Development:

The dietary management of the inherited metabolic disease Phenylketonuria (PKU) must count as one of the most clear and remarkable successes of clinical nutrition. Tens of thousands of lives have been lifted out of potential ruin by the use of special supplements within a strict dietary regime.

Obviously many people: scientist, dietitians and clinicians from doctors Garrod and Folling onwards have contributed to this success, and, as the Folling episode illustrates, the important role of the determined parent who won't give up was demonstrated on the very first recorded case of PKU. The medical history of PKU is well covered elsewhere and for this piece, as we celebrate the 60th anniversary of dietary treatment of PKU, I would like to highlight one narrow aspect that I have been closely involved in for 30 years - the development and technology of the special supplements used in the management of PKU.

To start at the beginning; a preliminary communication to the Lancet in October 1953 by Dr. Horst Bickel, a young German doctor studying for his PhD in biochemistry at Birmingham Children's Hospital in England, Paediatrician Dr. John W. Gerrard, who ran a clinic for children with mental retardation of unknown cause and biochemist Dr. Evelyn Hickmans, who ran the hospital lab. The article called "Influence of phenylalanine intake on phenylketonuria" paints a vivid picture.

Bickel had brought to Birmingham knowledge of Folling's ferric chloride test to diagnose PKU. With the approval of Gerrard, he applied this test to a group of mentally retarded children of unknown diagnosis. The third child tested, in April 1951, was Sheila Jones, and so she was diagnosed at the age of 17 months. Her mother, Mary Jones, was extremely grateful to learn the cause of her child's disorder but she immediately asked what the Birmingham team was going to do to help Sheila. This is an example of the gap between science and health care. The mother appeared in the hospital every day for six months persistently asking what they had thought of to help Sheila.

Although many of the products included in the kits were donated or discounted it still cost approximately $3,000 to assemble the 60 kits and ship them to various locations.

"We are really hoping that someone with the resources to sponsor this project sees the value," says Pallone. She also says that a little bit more funding could be used to translate some of the documents into French for the families who would prefer it.

The value has been noted by many since distribution of the kits in December.

"Thank you so much for the starter kit! Our little guy was born on Dec 19, and diagnosed with PKU on the 30th. Needless to say, it's been pretty overwhelming (especially for new parents), but the kit has made the transition smoother - both emotionally and financially."

~ Lisa Olsen, PKU Mom, Ontario

"The PKU Starter Kits are GREAT and will be a very special and meaningful intro to the PKU community!"

~ Barb Cheng, Dietitian, BC Children's Hospital

"The kits are awesome! To be able to give new parents a scale and the new Virginia Schuett book is priceless!"

~ Genevieve Lafrance, Dietitian, Sherbrooke, QC

If you are interested in supporting

At the age of 25 months Sheila Jones, - described as "an idiot" who spent her time groaning and banging her head - was treated with an experimental phenylalanine free formula, starting November 27, 1951. We know this thanks to the case notes of Dr. Gerrard. After she lost weight in the first weeks of treatment, the formula was changed to be low in Phe, starting December 18, 1951. As a side note; Louis Woolf, who was a biochemist at the Hospital for Sick Children on Great Ormand Street in London, England developed this formula concept. With a colleague he published the idea for this formula in 1951 because the doctors at his hospital refused to use it telling him to stop wasting his time on an obviously untreatable condition. Over the next year Sheila's symptoms and biochemistry greatly improved. In order to be certain that the improvement was due to the diet the Birmingham team began to add 5g per day of phenylalanine to the formula (a dose that today is more than the allowable amount in most PKU adults). The team did not tell the mother "so that any change could be noted without bias"! Once on the Phe rich diet the changes were drastic and disheartening. Sheila immediately exhibited many signs of regression including "banging her head, long periods of crying, inability to stand/crawl and periods of lifelessness". After hospitalizing the child they repeated this cycle with the same results and concluded "In this child at least, the beneficial effects of a low phenylalanine diet seem unequivocal".

*******The Birmingham team made a silent movie of the treatment of Sheila Jones. Gerrard gave his copy to Dr. Harvey Levy of Harvard University, who posted it on YouTube. It can be accessed on the CanPKU website. There is a second version of the movie with a voiceover by Bickel which can be obtained from Canadian PKU and Allied Disorders Inc.********

Dr. Bickel and others including Louis Woolf went on to try the diet in younger children with great success and laid the foundation for current screening and treatment. Bickel helped build a major centre for IEM research and treatment at Heidelburg, Germany.

In those days amino acids were quite exotic chemicals, only available at huge cost from specialist suppliers. They were analysed by means of paper chromatography, which Bickel had gone to Birmingham to study, using highly carcinogenic solvents. The sample was put onto the paper and, as the solvents flowed across it each amino acid travelled at a

this worthwhile project, please ***email Nicole Pallone.***

Hat's Off To...

The Provincial Government of Saskatchewan

As of January 1st, 2012 all residents of Saskatchewan living with an IEM (inborn error of metabolism) such as PKU receive coverage for the necessary low-phe metabolic foods.

This is a huge step for a province that previously had no support at all for this very expensive and necessary element of PKU treatment.

The coverage is somewhat limited in what products are available but is universal in that all patients receive assistance regardless of age, income or tolerance.

Saskatchewan resident Jodi Hoover is very happy with the new coverage. "We are so happy that we can now provide Brett with his special food and we don't have to limit him or exclude items because of the cost," says Hoover. "We ordered some items that we normally would do without like Camburgers and buns. Going through the boxes was a little like Christmas morning. What a gift! We are so very grateful!!"

On behalf of all the Saskatchewan families who are benefiting from this new funding we would like to sincerely thank the many families who wrote letters, arranged meetings and helped convince

different speed and were therefor separated. The paper was dried and then reacted with ninhydrin (yet another carcinogen!), the phenylalanine ended up as a separate blue spot on the paper which was cut out and mashed into a test tube with alcohol and the resultant colour density read in a spectrophotometer. So the idea of manipulating amino acids in the diet was quite a difficult and cutting edge concept at the time.

The formula used in these early experiments was produced by breaking a protein down into its constituent amino acids and then removing the phenylalanine. The following method was used: a protein such as casein was hydrolysed into its constituent amino acids by boiling it for several hours with concentrated hydrochloric or sulphuric acid (a dangerous and very odorous process) resulting in a thick, acid tar containing separate amino acids. This was then neutralised with a base such as sodium hydroxide and then clarified by addition of activated carbon and filtering, which produced a clear solution of amino acids.

This amino acid soup contained phenylalanine which was then removed by addition of another type of activated carbon, one with a strong affinity for aromatic molecules of which phenylalanine was one. This also removed the other aromatic amino acids tyrosine and tryptophan which then had to be added back. The result was a mixture of amino acids free (almost) of phenylalanine, and this was the basis for the Birmingham experiments.

This technology was developed commercially in Europe (SHS Ltd.) and the US (Mead Johnson) with such products as Albumaid XP and Lophenalac, and was the basis for PKU formulas up to the late sixties. The technology was refined with the use of ion exchange resins to replace the sodium hydroxide and carbon chromatography columns to help the separation of phenylalanine but basically it remained the same as that used by Bickel. The products worked very well but, as anyone around at the time will remember, they had a distinctive taste and smell! These products, though they seem primitive now were, in fact, quite pioneering and were probably the first synthetic or elemental diets long before the "space diets" that were developed for NASA in the sixties.

At the same time the analysis of amino acids developed rapidly with the use of ion exchange chromatography to replace the paper methods, and automated commercial instruments began to appear though they were unreliable -

the government to implement this great program!

News from BioMarin:

You may recall that we had previously announced delays in the development of our home blood Phe monitor program due to issues with accuracy at the low end of the Phe concentration range. Based on the results of the pilot clinical (donor) study, we have discontinued plans to develop the Phe monitor. The purpose of pilot clinical trial was to explore the limits of accuracy when testing the device with whole blood from individuals with PKU. And the results were much different than we expected. The inaccuracies extended well beyond even the range that we anticipated to be problematic with the Phe monitor.

It is unfortunate but we have come to the conclusion that the enzyme/strip technology that works so well for glucose monitors simply cannot achieve the accuracy that is needed to insure safe and reliable readouts for individuals with PKU. The approach has proven not to be transferable to the home Phe monitor effort. We have now brought this program to a halt. There are other early stage technologies being developed that may be adaptable to a Phe monitor and we will monitor progress on those efforts but at least for now, work with the current approach to developing the monitor will stop.

Thank you for your attention.

requiring skill and constant attention.

Towards the end of the sixties the Japanese developed the ability to manufacture very pure crystalline amino acids by the use of fermentation using special strains of bacteria that exuded amino acids as they grew. These pure amino acids were produced as pharmaceuticals for intravenous use but soon became available at a quality and price that made their use in special formulas possible and companies began substituting mixtures of crystalline amino acids for hydrolysates in PKU formulas.

This conferred several advantages; the amino acid profile could be adjusted to give a better nutritional value, the taste, though still pretty grim, was much better, or at least different from the hydrolysates. Perhaps most importantly it became possible to easily develop formulas for the other, rarer inborn errors of metabolism (IEM) such as maple syrup urine disease (MSUD) and Homocystinuria, which were almost impossible using hydrolysates, opening up many new possibilities for dietary management. This eventually paved the path for expansion into Tyrosenemia, Uria Cycle disorders, Methomelonic Aciduria, Proprionic Acidemia and others.

Over the following 40 years these pure amino acids formed the basis of most formulas used to manage IEMs though, during this time, the formulas have evolved significantly and many improvements in nutrition, taste and convenience have taken place, some of which I mention below.

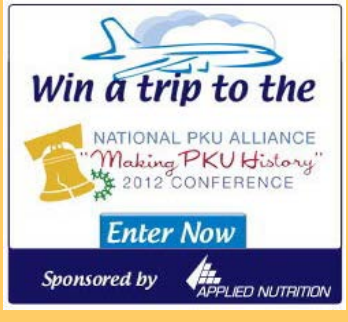
- Nutrition - This has improved over the years as knowledge and technology advanced, formulas for IEM's have been on the leading edge of human nutrition, for example they pioneered the use of trace elements such as selenium and cobalt in human diets and were among the first to investigate and use long chain poly unsaturated fatty acids such as DHA and arachidonic acid using state of the art raw materials.

- Taste - Amino acids are as varied in taste as they are in chemistry from sweet and pleasant (glycine) to tasting like burnt feathers (tryptophan) and boiled cabbage (methionine). When mixed together the result is a horrible mixture of sweet, sour, bitter and so on. Manufacturers have learned many tricks and techniques to improve on this to the point where the taste of a standard amino mix is not too bad (I may be biased on that one!)

Elaina Jurecki, MS, RD
Associate Director,
Medical Affairs

Making PKU History with the National PKU Alliance

The National PKU Alliance hopes you will join them for their 2012 Making PKU History National Conference being held July 26-29, 2012 in Cherry Hill, NJ. The conference promises the latest on PKU topics, such as hepatocyte liver cell transplantation, gene therapy, PEG-PAL, PKU management tips, neurocognitive issues, maternal PKU and more presented by the most innovative leaders of the PKU community. In addition to learning about PKU research, advocacy and education, you will also have the opportunity to network with other individuals living with PKU.



The conference will include several new features such as the PKU Adult and Guest Dinner, the Kids Zone and a Scientific Poster Session! The NPKUA will also honor their 2012 PKU Heroes and PKU Pioneer at the conference. **Click here** to learn more and start planning your trip! Here you will be able to register, learn how to book your hotel room, and view the full conference agenda. The NPKUA is also offering partial scholarships to adults and families that need financial support to attend the conference.

This is the conference you don't want to miss! The NPKUA is excited to offer several promotions:

- Early Bird Registration - Register before April 30, 2012 and receive discounted
- Convenience - The formulas were originally presented in baby food cans; PKU was, in the early days, a paediatric condition. But with the success of the dietary management the population has gradually grown and aged, the form of the formulas has followed this trend. Starting with pre weighed and flavoured sachets that just need water addition, and then liquids and semi solid weaning formulas in dosage packs. Manufacturers such as Vitaflo and Nutricia have developed forms of the products which look like the standard commercial drinks used by the peers of teenage patients with the hope of normalising the taking of the protein substitutes.

The gradual improvements of the kind mentioned above have, I think, greatly assisted the nutritional management of PKU. In the early days of hydrolysates and the initial amino based formulas the main problem was overcoming the effect of poor taste and large volumes of the protein substitutes. I believe that the current generation of liquid and semi-solid formulas with small volume and much improved taste have ceased to be the main problem in a typical PKU regime. It is unfortunate that in many academic papers it is routine to refer to the bad taste and poor acceptability of these products, I believe this thinking to be a bit behind-the-times and that the main challenges in the successful management of PKU and other IEM's lie elsewhere.

There have been other developments in special formulas in PKU using difference approaches, for example:

- Large Neutral Amino Acid (LNAA) formulas - Formulas based on this approach were suggested in 1985 by Professor Lou at the Kennedy Centre in Copenhagen; they have been used successfully but not extensively to manage PKU and in theory offer the prospect of a more relaxed dietary regime. They are based on the idea that phenylalanine and the other LNAA cross the blood brain barrier and the gut epithelia using a common transport mechanism and hence compete with each other; therefore supplementing a perhaps slightly relaxed diet with LNAA will cut down the amount of phenylalanine that can be transported into the brain because of the extra competition. The technology of such products is the same as conventional products, consisting of amino acid mixes in powder or tablet form.

- Glycomacropeptide (GMP) - GMP is a peptide that is

prices!

- NPKUA Membership Discount - Become a member of the NPKUA for $25 and receive discounted registration rates!
- Registration Refund Contest - Who will attend for free? One person out of the first 100 registrations will get a refund on their registration!

The National PKU Alliance hopes you will join them in Making PKU History with their 2012 National Conference!

naturally low in phenylalanine. It is a by-product of cheese production and is now available commercially. The material is soluble and tastes like casein (stale milk); current material has about 200mg of phenylalanine per 100g. This makes it attractive as the basis for protein substitutes as it offers the possibility of a wider variety of better tasting supplements, but there are some nutritional issues that need to be addressed. The amino acid profile of GMP is nutritionally extremely poor and hence needs supplementing with extra essential amino acids to make it nutritionally acceptable, this adversely affects the taste and so there is a trade-off between good taste or good nutrition. GMP products have been available for many years in Europe and more recently in the US; controversy exists as to how much the nutritional value needs to be improved to make an acceptable product for long term management.

The future:

It is obvious that there will be many changes in IEM management as medicine and biotechnology rapidly advance. In PKU for example an enzyme cofactor BH4, is widely available, enzyme substitution is on its way and genetic manipulation is advancing in animal models. Amino acid formulas continue to be instrumental in the management of PKU as new treatments are developed since it is by far the least costly option and they will continue to be an important tool until a cure is developed. Technology is still advancing the way products are produced, helping their palatability, textures and nutrition.

One thing is certain and that is that the rapid change that has taken place in the management of PKU and other inborn errors of metabolism will continue - some things never change!

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.

Sincerely,
Canadian PKU & Allied Disorders Inc.

Because Knowledge Leads to Better Health

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