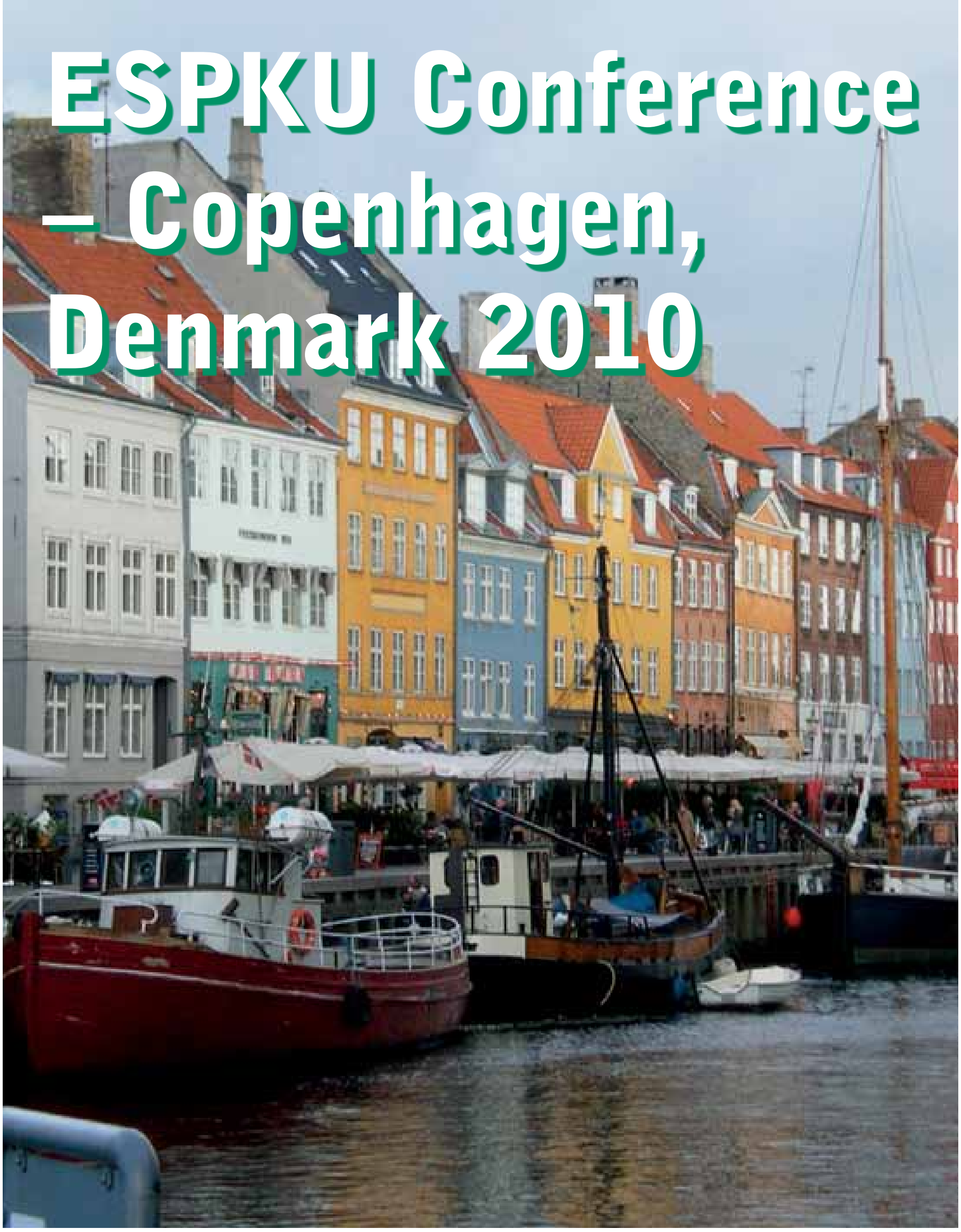
NSPKU
News&Views
Winter 2010/2011
The National Society for Phenylketonuria (UK) Ltd
Issue 133

Inside this issue...

ESPKU Conference – Copenhagen, Denmark 2010



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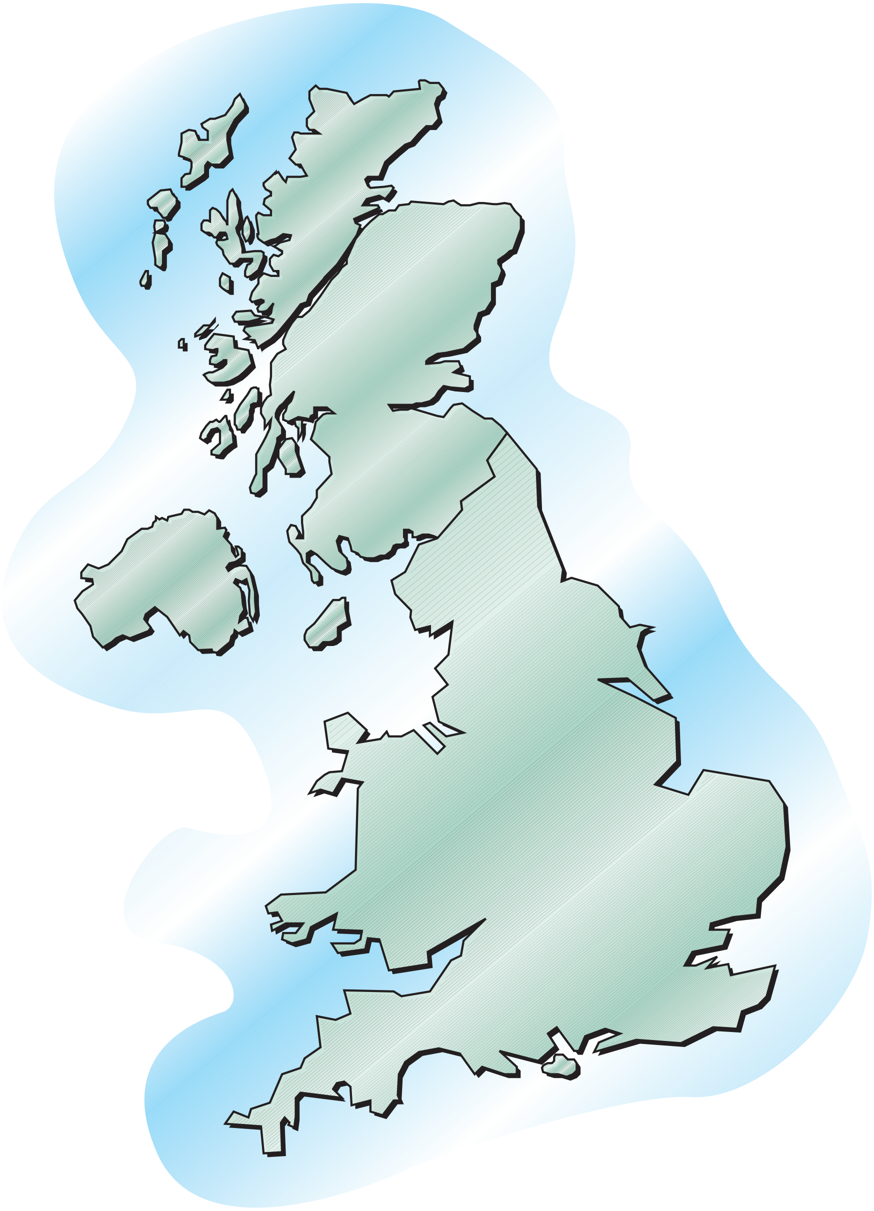

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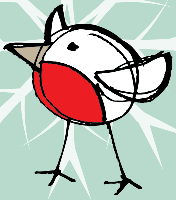

Editor's Report...

Hi Everyone

Well winter has arrived with a vengeance hasn't it! It's time to start filling those freezers with warming soups and stews. I have included a couple of Kylie's favourites for you to try.

The teen page is looking for articles from our young people, I am sure there are lots of you doing interesting stuff all the time that readers would love to know about. Please send your contributions to me.

Enjoy Christmas and I look forward to receiving your stories about the festive get togethers for the next issue.

Ed

NSPKU
The National Society for Phenylketonuria (United Kingdom) Limited

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Letters & Special Thanks

News & Views, 23 Barnfield Cottages, Arundel Road,
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Email: editor@nspku.org

Dear News and Views,

My name is Cameron Hamer, I am 8 years old and have PKU. In September it was my Granddad's (Michael Whitaker)70th Birthday. He had an open house but did not want any presents so we put some donation boxes out for people to put in what they liked. We collected £400 and enclose cheques totalling this amount.

Even though I am only 8 years old I know what I am allowed to eat and drink and I am very good with my exchanges. My Dieticians are really pleased with how I deal with and manage my condition and my levels are always spot on.

I would like it if you could please print this letter and the picture enclosed in the quarterly magazine so we can show it to the people who kindly donated the the NSPKU as most of them had never heard about it.

Thank you very much,
Cameron Hamer

Dear News and Views,

My name is Scott Rae, I am 6 years old and have PKU. My Uncle Paul had a 40th birthday party and people gave him money instead of presents. I have sent a photo in of myUuncle Paul giving me a cheque for £450 which I have given to the NSPKU. I think it was really good of Uncle Paul to give this donation and thank everyone who went to his party and gave money. I was too young to go but my Mum and Dad say it was a really good party.

Best Wishes
Scott Rae

Dear News and Views,

I enclose a cheque for £410 for the NSPKU. £210 of which is the remainder of the monies collected as a result of Arwyn's "Offa Dyke" walk last May, and £200 collected by a local choir, Sain Teilio, of which Arwyn, myself and Wendy, our daughter, are members. We sang carols at our local town a Saturday before Christmas.

Yours sincerely
Arwyn Davies

Dear News and Views,

My husband and I got married in York on 7th May 2010 and here is a donation from the money we received from our guests. Enclosed is a cheque for £200.

Our daughter, Eve, has PKU and is doing really well. She is 18 months old and is a typical toddler! Her and her brother, Ellis, made our wedding a wonderful day, and all our guests said how amazing and beautiful they were on the day.

We would like to thank the caterers, Bradshaws of York, for bending over backwards to cater for Eve and making it a hassle free day.

Thank you also to the NSPKU. The work you do is wonderful and we hope this donation will help to continue the support you give to the people with PKU and their families. It is not always easy caring for Eve, but reading the articles and letters in the News and Views comforts us, and we know we are not alone.

Thanks again
Helen, Andrew, Ellis and Eve Plant

Dear News and Views,

My grandson, Jordan Davies, suffers with the condition PKU and the members of the bingo club have kindly donated the raffle monies to this charity. I would like you to accept this cheque of £1000 on behalf of the Misty Waters Bingo members.

Yours truly
Linda Davies

Dear News and Views,

Our son, Matthew, is 18 months old and has PKU. In June Matthews daddy (centre) and his 2 uncles ran the Lisburn 10k to raise money for the NSPKU. They raised £535 and Matthew went along to support! Thank you for all the work you do as a charity to support families like ourselves.

Yours sincerely
David and Juanita Morrow

"Thanks to **Vicky Ellis** from **Huddersfield**, who regularly sends in cheques, the proceeds of an NSPKU collecting box where she works at Cohens Chemist. Since November last year the total sent in is **£75.07**. **Thank you Vicky.**"

The treatment of phenylketonuria varies for each individual patient. No patient should alter their own treatment as a result of reading how another patient manages their diet without first consulting their doctor or dietitian.

Dear News and Views,

I decided to raise money for the NSPKU as my best friend has two children who suffer from PKU. I wanted to raise the profile and money of the charity.

I challenged myself to take part in the British Gas, Great British London swim, which was to swim 1 mile in the London Royal Victoria Docks on 3 July. It was a fabulous day with over 5000 swimmers, a great atmosphere and a well organised event. I enclose my offline sponsorship of £736. I had set up a just giving page with online sponsorship of £214 giving a total of £950 raised. I enclose a copy of the newspaper article that was in our local paper before the event.

MAKING A SPLASH: Sara Jones (inset) from Johnstown in training at the Carmarthen Leisure Centre, ahead of her mile long swim in the Royal Victoria Dock in London raising money for NSPKU.

Swim inspired by children's illness

The family I was fundraising for are Aled, Wendy, Ifan and Awen davies of Llandeilo, Carmarthneshire

Many Thanks
Sara Jones

Dear News and Views,

Please find attached a cheque for £280. I've also attached my just giving page where I raised an additional £870 for the NSPKU. *http://www.justgiving.com/Helen-Pinfold*

My Nephew, Jack, has PKU and I wanted to show my support by raising money and awareness for the charity. I completed the "run to the beat" half marathon in October within 2hr 30 minutes which was good for me as I have never raced before! I have now entered the London marathon 2011 where I hope to raise even more money for such a good cause.

I have enclosed a photo of Jack which I would be grateful if you culd print with this letter.

Yours faithfully
Helen Pinfold, Bromley

Dear News and Views,

Almost exactly 10 years ago we had a "welcome to the world" party for our son Stan, who was born in July 2000, with PKU. It was a fantastic party and really cheered us up when we were still new PKU parents, and very unsure what life with a PKU child might be like. Now, 10 years on, we know that although it has its inconveniences there is really nothing to worry about. Stan has of course turned into a boy who is only different from "normal" children in good ways, and we are very proud of him.

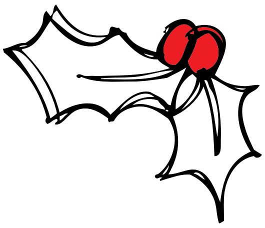
Just after the party our friend Christopher O'Reilly, a very good, though as yet unpublished. poet wrote a poem about it, and about Stan. Reading it again recently I was struck by what hope and encouragement it expressed and thought you might like to publish it in News & Views.

For Stan

What can we say, after drenching
your E.T. head in sparkling Cava
the afternoon blowing a light bubble
of chatter and laughter
across the sun's steep shadow
to burst into drowsy evening
drops of incoherence scattered in a fond explosion?

But that you, Stan -
sleeping through your own party -
are there as you
and how your world now is -
one fine scratch on your D.N.A record
meaning a whole different beat
through your own life's album -
a unique intro. to now follow through

Boy, who will only be
at all ordinary
to those that know you,
may you beat on through
to harmony,
to something true.
Anyone can say that,
but perhaps it's what you'll do.



Best wishes,
Richard Brook

News from the UK Newborn Screening Programme Centre

My name is Caroline Bridges and I have a 11 year old daughter, Hannah, with PKU. About seven years ago I answered an advert in CLIMB (The National Information Centre for Metabolic Diseases) magazine. They were looking for parents to help health professionals set guidelines for the PKU screening process. Since then I have sat in on various **UK Newborn Screening** meetings as a parent representative. I have used my PKU screening experience to give health professionals' guidance when developing and implementing PKU screening standards. In 2005 the **UK Newborn Screening Programme Centre** set standards for PKU screening to *ensure timeliness of the process, population uptake and timely diagnosis, assessment and treatment of babies screened positive.

(taken from the Executive Summary)*

In the 2005 guidance from the **UK Newborn Screening Programme Centre** it was acknowledged that some laboratories use different thresholds and methods for the measurement of phenylalanine. Since 2009 all laboratories have introduced Tandem Mass Spectrometry as the screening technology for PKU. In addition there have been developments in knowledge about biopterin disorders and a need to be consistent with standards and guidance for other screening programmes and therefore a need to revisit the protocols. The PKU Expert Working Group consisting of representatives of the **UK Newborn Screening Programme Centre, British Inherited Metabolic Disease Group, UK Newborn Screening Laboratory Network** and the **National Metabolic Biochemistry Network** has been carrying out this review for the past two years. The revised standards/guidance for PKU screening and the **PKU Expert Group Report** has now been posted on the UK Newborn Screening Programme Centre website **www.newbornbloodspot.screening.nhs.uk** and these standards will be implemented by 1st April 2011.

Your Screening Stories

I vividly remember being told by my GP that Hannah had PKU. I remember it was a Friday and we weren't given an appointment with the Consultant until the following Monday. I remember my GP not really knowing anything about PKU, apart from what he had just quickly looked up in his old student notes. I remember what a heartbreaking weekend we had until the Monday morning when we saw the Consultant and Dietician. I'm sure all of us have our own screening story and although we eventually saw a brilliant consultant and dietician the lead up to that was not so good. (The Midwife said to me when she came to take Hannah's heel prick test "It's only routine and it won't affect you").

To support The UK Newborn Programme Centre's important work the Centre would like to hear your screening story, good or bad. This information will be used to check standards across the UK are being met and to improve in areas where they aren't.

Please email your screening story (including, if possible, the following points) to **christine.cavanagh@nhs.net** or phone **Christine on 07786 313100**

- Date and location of birth?
- How old was your baby when He/She had the heel prick test?
- What information were you given by the midwife?
- How old was your baby when you were told that He/She had PKU?
- How were you told and who told you?
- When did you have your first appointment and who with?
- How were you treated throughout the process?

THANK YOU

A Publication is Born

The now produces a really large selection of leaflets and booklets which are available on our web site and in print. There are well over 20 of them and at each medical meeting we discuss which need updating and ask if there is a need for any other subject to be covered. We nearly always find that there is and wondered why we hadn't appreciated the need earlier.

The Medical Advisory Panel (MAP) of the NSPKU meets twice a year, usually in Birmingham, in May and November. A dietitian, doctor or NSPKU committee member will come up with a suggestion which we discuss. It's often when someone has had a problem and needed some guidance which isn't written down which sparks the discussion. Writing leaflets and booklets is quite a difficult and time consuming task but in the famous words of the philosopher Plato "The beginning is the most important part of the work". One of the committee will volunteer to take the leaflet on and write the first draft and 'bones' of it. This start is a huge leap forward and it is then circulated to all the MAP members for their comment. They are given about a month to get back to Lucy, our administrator, with their views. Lucy will then pass these on to the author. The author will incorporate the views and advice and a second draft goes round. It is surprising and helpful to get all the heads together in this way as everyone has their own expertise and different ideas about how advice is expressed. Sometimes a sentence can be taken in quite the opposite way in which you intend. An example of this was in the re-write of the dental leaflet. We wanted to advise about the dangers of eating too many sweets between meals so we started by saying 'Eat sweets at mealtimes'. This could be interpreted as an order meaning 'sweets must be eaten at all meal times' instead of what we really wanted to advise i.e. sweets are less harmful to the teeth when given at mealtimes.

It can sometimes take several drafts to be circulated for everyone to be happy and sometimes you just have to call a stop as changes are made to changes; it can go on forever!

The art work is really important as we want leaflets to be read. One of my favourites is the Packed Meals for a PKU diet. It's so attractive and encouraging I want to eat all those things on the cover. All new publications are sent out to members of the Society with their News and Views so that uses up about 1000. The more you print the cheaper it becomes so there is always the temptation to print too many. You think at the time 'that's it, this will last a long time' but actually life and times change so it's false economy to over-do the numbers and storing takes up a lot of space and paper is heavy.

So, have you ever thought there is a gap in the topics covered by our leaflets? Let us know about it and it can go before the MAP – you might even be asked to do a first draft for us but don't let that put you off. Remember Plato, you could get the wheels in motion.

Eleanor Weetch, Society Dietitian

MR PROMIN'S LUNCH BOX LOGISTICS

MON	TUE	WED	THUR	FRI	SAT	SUN
✓	✓	✓	✓	✓	X	X

5 DAYS X 7 GREAT FLAVOURS = 2 DAYS OFF!

SUM TOTAL = MORE THAN ENOUGH SNAX & CAKES TO FILL A WEEK OF LUNCH BOXES

NOW AVAILABLE ON PRESCRIPTION

SNAX

ARE AVAILABLE IN FOUR FANTASTIC FLAVOURS

CHEESE AND ONION READY SALTED
SALT AND VINEGAR JALAPENO

AND TARANIS CAKES ARE AVAILABLE IN 3 SCRUMPTIOUS FLAVOURS

TARANIS LEMON, TARANIS PEAR & TARANIS APRICOT

PROMIN

338 TURNCROFT LANE, OFFERTON, STOCKPORT, CHESHIRE. SKI 4BP
TEL:(44) 0161 474 7576 FAX:(44)0161 474 7576 E-MAIL: FIRSTPLAYDF@SMARTONE.CO.UK WEB: WWW.PROMIN-PKU.COM

Because they really do taste as good as they look...












All recipes developed, tried and tested by Eileen Green using:

Fate Low Protein All-Purpose Mix
Fate Low Protein Cake Mix
Fate Low Protein Chocolate Cake Mix

FATE
Special Foods
TM

Ask your doctor to prescribe Fate mixes, then ask your chemist to telephone our direct order line : 01215 22 44 33. Helpline and general enquiries tel: 01215 22 44 34

Fate Special Foods recipes

Hello

I can't believe it is almost Christmas time again! This is a lovely recipe to make anytime, especially at Christmas. I use a mixture of ground ginger and mixed spice which gives a good flavour. It is important to measure the golden syrup accurately in this recipe. A metal spoon dipped in very hot water will make it easier, as the syrup will just slide off it.

The most common failure in gingerbread men is that they spread when put into the oven, so make sure you do a test bake. This recipe is enough for about 10, depending on the thickness you like them.

Ingredients

300g Fate All-Purpose Mix

100g block margarine

100g soft light brown sugar

1tsp ground ginger

1tsp ground mixed spice

90g golden syrup (the old fashioned kind in a tin, do not use pouring syrup)

15mls (1tbsp) water

few currants and glace cherries for decoration

FATE GINGERBREAD MEN

Method

Pre~heat oven Gas 6 200°C 400°F

Place the Fate All-Purpose Mix into a bowl, and rub in the margarine until it resembles breadcrumbs. Stir in the sugar and ginger. Add the golden syrup and water, and mix with a metal spoon until the mixture forms a dough.

At this stage you may begin to think that the mixture is too dry, but do not be tempted to add more water or syrup, just keep mixing and it will all come together. Lightly dust the surface with extra All-Purpose Mix, and knead the dough a couple of times until it is smooth and not crumbly at all.

Lightly dust the surface again with All-Purpose Mix and roll the dough out to about ½ inch thick. Cut a small piece of dough and place in the oven for a test bake. If the biscuit mixture runs, add more Fate All-Purpose Mix to the mixture and try again. If successful, carry on! Cut the dough into shape with a cutter. Push currants into the dough for eyes and buttons, and use small pieces of glace cherry for the mouth. Place on a lightly greased baking tray and bake for 10-15 mins until golden brown. Leave to cool on the tray for a few minutes as they are very soft when hot, then transfer to a cooling rack.

Note: Instead of gingerbread men you can use the same recipe for other shapes. When the girls were little, we would use this recipe to make Christmas tree decorations at Christmas time. Cut the dough using Christmas cutters, such as Christmas trees, stars, and angels, place on a lightly greased baking tray and put in the oven. After 10 minutes of baking, use a straw to make a hole in the biscuit and return the biscuits back in the oven. After baking and cooling, it is nice to ice them and use pretty ribbon to hang them from the Christmas tree.

They keep well in an airtight tin, or they can be frozen.

Teen ZONE

These pages are for young people to have their say. Send us your stories and we will give them Pride of place here!

Astrology Word Search

Words can go horizontally, vertically and diaginally in all eight directions. Words may overlap but do not share letters.

J R R K P R H Y X V V C N H L W F N C W X B K L J
Z R L V J R S V N I Q M P N Y G E M I N I C N X L
F R D S J C K U R O H C Z Z F G H N H L R V K T M
N Z M E B T L G I L I N R B F M L N N B P Q F N P
H W K I L V O R R R C T R K Q V E P I S C E S A R
G H L R B G R Y F O A I I P Z P L N N H R M M D T
W N C A L M K K N T L T R S T G V C L K T M G N X
R O A L G J K J T O R T T U O L A R B I L D T E K
K O J S W C U T C Q P N N I H P K N L N N N G C J
D M J M P N K M M L X E F T G T P L K R B C W A R
D C L G C E U M Z F S M F W L A B O L O X P N M B
R P H T Z I C B K U X N K L M H S R T C H J Q G J
T P I V M L N T R H M A R S C M C V K I W Q R Y M
Y O N M C K K U S T S M N M D V J N N R P Q Y H G
N T I C N R A Y V L S A J O T U L P R P R K L N K
K X P H Q T G L P U K Z T B P W J T N A D T T M Z
X C V V T R K Y I W M Z M U J P D N Z C L L R N C
C K A M F L M R F J K Y L T R N B Z W J U R L N Q
V C M N W K A K J H R S V M T N U Y Q X R K E N V
J Q L W C U X F Z U D E L J N Q G S W G A J O F H
Y R V L Q E W X C R N X H K W B P B R V N M R P R
X Q K A H K R R Y D Q T T F L N F W T G U W Z N J
R H K F T W E L D J R I R E N I R T L F S T N R C
J M V Y P M K F Q X H L M M M D J L P K G F N T R
D R T D N T F G B W Q E P Q F N F K L Z K X L H D

acendant
aquarius
aries
aspects
cancer
capricorn
conjunction

gemini
immium coli
leo
libra
mars
mercury
moon

neptune
opposition
pisces
pluto
sagittarius
saturn
sextile

sun
taurus
trine
uranus
virgo

juvela
low protein

The Juvela Low Protein range of foods has been specially developed for those on a low protein diet

Our range includes a Sliced Loaf, Bread Rolls, All Purpose Flour Mix, a selection of Cookies in Cinnamon, Orange and Chocolate Chip flavour; and a Pizza Base complete the range. (Clockwise from above)

From May 2010, the responsibility for supplying the Juvela Low Protein product range switched from SHS Nutricia to us at Juvela. You will still be able to obtain these products on prescription.

What's New…

Dedicated Customer Support Services:

New!

☎ UK/NI Free Phone: 0800 783 1992 • ROI Free Phone: 1 800 40 50 90

Unit 8, Juvela (Low Protein), 19 De Havilland Drive, Liverpool, L24 8RN

@ lowprotein@juvela.co.uk

Try Something New…

Keep in touch with us via Email, Post or Phone if you would like to receive free samples of any Juvela products, or a free copy of our new recipe book!

New!

'Make Each Meal An Opportunity'

Juvela Low Protein Product Guide and Basic Recipes

Coming Soon…

www register now at:

www.lowproteinfood.co.uk

New website Coming soon!

UK freephone 0800 783 1992
ROI freephone 1 800 40 50 90
lowprotein@juvela.co.uk
Juvela, 19 De Havilland Drive, Liverpool L24 8RN

Crunchy Mango Layer

Delicious treat in a matter of minutes!

Ingredients

1/2 mango, diced

5 JUVELA Low Protein Cookies (any of your favourite flavour)

Lemon/Mango Sorbet* to serve
(*consult the NSPKU guidelines for suitable sorbet)

Method

1. Place the Cookies in a plastic bag and crush using a rolling pin.
2. Assemble the dessert in a tall glass by spooning in 2 tablespoons of Cookie crumbs, 2 tablespoons of mango, follow by spooning 2 tablespoons of sorbet and finish off with the remaining Cookie crumbs.

ESPKU Conference 2010 Copenhagen Denmark

I found myself in the lucky position of standing in for Eleanor in an autumnal Copenhagen last week. I had a really stimulating and fun three days, thank you NSPKU and here's my feedback:

ESPKU now has online conference registration so we know exactly who was at the conference. There were 3 programmes running at the same time – one for the representatives of different National PKU societies, one for professionals (which I'm reporting) and one for people with PKU and their families. (We had some shared sessions as well).

The conference goers were:

- 68 people with PKU
- 110 relatives of people with PKU
- 70 professionals
- 19 "delegates" from member associations
- 60 representatives from industry

Throughout the conference there was an atmosphere of warmth, friendship and ideas. The Danish hosts had organised a fantastic array of events for children – time on the beach, on a nature reserve, cake decorating, a disco, balloon animal making – to name a few. At each mealtime the PKU buffet was an array of beautifully presented food with plenty to choose from at all times of day.

Misfolded Molecules?

Ania Muntau showed us some diagrams of how the Phenylalanine hydroxylase enzyme is folded and how people with PKU may have "misfolded" enzymes that stop them working. BH_4 may help restore the correct folding (and therefore the correct shape – which is crucial to enzymes) to a misfolded enzyme molecule and this is how sapropterin (Kuvan) works. Prof Muntau did tell us that there are other potential PKU treatments possibly in the pipeline, but they are several years away. These possible treatments are: enzyme substitution therapy – called PEG-PAL – which could be injected weekly; also liver cell transplantation; and finally gene therapy to the liver – this would be targeted delivery using "intelligent particles" and could even involve iron particles and magnets. BH_4 is the treatment that is in use in several centres in Europe and other speakers went on to explain how it is used.

Dutch dietitian
Reet van Rijn

How do we test to see if BH_4 can be used?

Amaya Belanger from Madrid told us about the 250 people in her centre with PKU – they range from 12 days old to 40 years old and BH_4 treatment has been used in Madrid for 8 years with 80 people taking it. The Spanish PKU population is 1500 strong, and a high proportion has the milder PKU which is more likely to respond to BH_4. Amaya and her colleagues have developed some sophisticated BH_4 response tests. More questions are arising now – once we know if someone responds to BH_4, can we predict how much additional protein might be tolerated? What are the proven and fully studied benefits of BH_4 and what are the challenges for maintaining a balanced and healthy diet whilst taking BH_4? More detail was later provided by the Dutch dietitian Reet van Rijn when she outlined the Groningen approach to starting on BH_4. Dietary tolerance is gradually worked out in a logical and step wise manner of recording food intake and measuring Phe whilst adjusting BH_4 doses, which can take many months but gives clear answers for all involved.

More Questions than Answers?!

The UK's own Anita MacDonald gave us plenty of food for thought – there is a lot more work to do on many areas of the PKU diet. Anita called for us to collect detailed information on Phe tolerance in those over 10 years old and to study what the causes of obesity in PKU could be (is it eating more calories or doing less physical activity, or both?) More information is needed on the nutritional needs of children and adolescents with PKU, and fibre in the PKU diet should be considered. There is scope for more product development, such as thinking about functional food components like prebiotics and whether they could be incorporated into protein substitutes.

To make life easier, how about a "one stop shop" for the 200-300 different specialist products that are now available to people with PKU…..and how about product development of more normal looking convenience foods since PKU families and adults have busy lives too. These are just a few of Anita's questions – she inspired us to consider all aspects of the undiscovered PKU world. Watch this space as more research gets underway.

Copenhagen's New Harbour or Nyhaven

Thor with a few of the 60 PKU conference go-ers

What really happens when Phe levels rise…. reports on children, adults and their families?

Thinking clearly and quickly?

In Poland Dr Bik-Multanowski used a special set of cognitive (thought process) tests called CANTAB. The tests looked at higher "executive functions"; these functions are thought processes that enable flexible adaptation of thought and behaviour in response to changing needs in a person's surroundings. Dr Bik-Multanowski tested 50 adults with PKU and those with high Phe did less well in their cognitive tests than those who were well controlled. Could this help people think about and discuss their diet more readily?

Mood Swings?

Dutch PKU families took part in two fascinating projects which may confirm what many of you already suspect. Firstly Karen Anjema from Professor van Spronsen's research team told us how she made over 500 phone calls to 51 children and young adults with PKU and their families or partners to ask if they can predict when their Phe levels are high, and ask what behaviour is like with high Phe levels. Mood swings and introvert behaviour were both shown to be associated with higher Phe levels. An impressive 86% of the patients themselves were able to correctly predict when their Phe levels were above the recommended level. (But…..people with higher Phe levels underestimated their levels and those with lower Phe levels overestimated levels).

High levels of Phe cause fatigue and low mood:

Amber ten Hoedt reported an elegant study of 9 adults with PKU who were well controlled on diet but who deliberately took in extra Phe and measured the results. Neither the participants nor Amber knew when they were having the additional Phe as Amber's study was a double blind cross over trial – 4 weeks were spent taking placebo or additional Phe, then a break or "washout" period for 4 weeks, and then a swap over to the other "treatment" of placebo or Phe. In the placebo period everyone's Phe levels were under 600, in the extra Phe period Phe levels were over 1200. Mood on extra Phe was low – participants felt less vigorous and more fatigued and depressed. Relatives and partners backed this up; and Amber also showed in thought process testing that slower reaction times and slower recognition of complex patterns occur with higher Phe.

PKU helped me become an Ice Hockey Star says Thor:

A real conference highlight was near to the end when Danish ice hockey star Thor Dressler (from the Hilvedore Wolves) told his story. Thor wanted to be a sportsman from early on – he loved hockey at age 5. Through his teenage years sport was popular amongst all Thor's friends who would also have liked a career in sport. Only Thor has made it – this is because his PKU meant he understood "right and wrong" from making choices with his food, from a very early age. He has continued to be focused on his sporting goals during his teens – whilst his friends were distracted by alcohol and partying. Thor is now 31 and is at the top of his game – he brought a signed shirt and happily posed for photos! He really was inspiring and said again and again that PKU didn't stop him, it helped him and it shouldn't stop others from achieving their goals. Thank you Thor!

I haven't mentioned fatty acids, nutritional follow up through different life stages, feeling full with GMP…….or the brilliant overviews of PKU from Harvey Levy and Francjan van Spronsen. I recommend to you all ESPKU 2011 in Poland.

Suzanne Ford, Senior Dietitian Bristol Hospitals

ESPKU Conference 2010

Eric Lange reports his experience of the Conference

I arrived at Copenhagen airport around 4pm on the Thursday…breezed through baggage reclaim … bought a ticket for the brand new M2 rail track to the hotel. Two stops. Alighted at Femeuren. Then off to find the Hotel. I asked a local where it was. .. he pointed behind me. Upon turning around I was greeted with a mass of scaffolding …renovation works were underway.

After registering I made my way to my room which was practical and modern with a lovely view of the bay…not like some unfortunates on the other side of the hotel who had scaffolding outside their rooms with little or no view.

I met old friends and made many new ones while picking up the programme of events.

In the evening the opening ceremony was at the Kennedy Centre in downtown Copenhagen, named after President Kennedy, this centre is a National Research & Counselling Centre for Genetics Visual Impairment and Mental Retardation. It is also the national centre for the dietary treatment of PKU. There is a website with a limited English translation - www.kennedy.dk

The first day began by the ESPKU Chairman David Abeln welcoming everybody and introducing our first speaker Prof Levy …a heavyweight in the PKU world of Research. He gave a wonderful overview of the history of PKU. After Prof Levy we were introduced to Prof Muntau of Munich who gave a talk on BH4 as an alternative treatment for mild PKU. Her full research paper can be found on the web here- http://www.biopku.org/pdf/muntau_NEJM.pdf

After that it was off to the delegates program.

The Delegates Program

This is dedicated to the delegates of each country.

Our program began with "story telling". This session as taken by Weber Shandwick. A very interesting seminar on how to tell a story and convey your message - with the emphasis on PKU.

It wasn't as easy as one thinks - especially if your background is not in the media or communication. We were shown the structure of a story with examples of each of the 6 steps… establish a solid message; be clear, creditable and persuasive; Identify your audience and the message you wish to convey.

It was interesting looking at examples of well-known people and how they "bend" the truth to convey the message. Branson and Clinton to name but two. Who'd be a politician with a conscience? Never believe what you hear and always do your own research.

Having been through the theory we divided into two groups to complete the first exercise based on what we had learnt. Very instructive. After lunch we conducted another exercise with interesting results. Both groups chose to create a story that helps budget holders understand the need for reimbursement for treatment of PKU.

Independently of each other, we came up with the same arguments and conclusions. Given the delegates come from all over Europe-it would appear that we all sing from the same song sheet and are all dedicated to argue for PKU patients-wherever they are.

What we were told is that in "batting" for PKU we need to grab attention ahead of others-because if we don't grab attention someone else will. So we really need to take heed of what we had learnt during the day.

Later on in the afternoon we had our "slow dating" session where we met other delegates and discussed at length our respective organisations and how we could help each other. Many emails and addresses were swopped.

We also had a presentation from a very inspirational mother from Turkey …Deniz Atakay (who organised the 2009 Conference in Turkey). Parents of PKU patients in Istanbul have organised a shop that specifically deals in baking product and cakes for PKU patients. It's a very social event and they all get to swop stories on coping with PKU.

We also had a presentation from the Austrian PKU society, which organised a music event by Musical Unplugged that included Garry Howard from the Flying Pickets. The event raised funds for their Society-OGAST.

Artistic endeavours

Icelandic PKU colleagues

In the evening we had the AGM. Points to note are as follows:-

1) ESPKU's policy is for all patients to have access to all treatment options-not limited by origin, age and genotype & include all treatment types.

2) The ESPKU petitioned the EU Parliament on food labelling –The ISA (The Independent Sweeteners Association) wanted to do away with the mandatory labelling of aspartame. Fortunately, the labelling will continue as is for the foreseeable future.

3) It has published a book "Living with PKU". Real stories from real people with PKU from all around Europe. Well worth a read. (This will be available at our Conference in March.)

4) The ESPKU applied to join Eurordis (European Rare Disease Society) http://www.eurordis.org/

5) The ESPKU website has been refurbished and is now multilingual.

6) Changes to the administration and organisation of future ESPKU Conferences to take effect immediately. The 2011 conference will be in Warsaw, Poland and 2012 in the UK.

7) The ESPKU conducted a European wide survey to understand the attitudes and behaviour of patients, families and healthcare professionals treating PKU. The aim is to identify gaps in service provision. An interesting outcome from this is that 75% of patients stray from the diet in one form or another.

The second day we joined the common programme, which along with the first day is documented in the "PKU Post" on pdf, files downloadable from the ESPKU website- espku.org.

However it is worth mentioning the look of joy on the children's faces when they were treated to a photo shoot with Thor Dressler –a Danish Ice Hockey International with PKU. Truly inspirational for the locals.

The excursion took us to the waterways of Copenhagen. A nice boat trip with a running commentary on the History of Copenhagen. However, imagine our disappointment when we were told that the Mermaid had been loaned to Shanghai for the 2010 Expo - see photo. Some of us stayed behind for an hour or so of window-shopping. Copenhagen is a beautiful city, very clean and the people are exceptionally friendly. Although quite expensive, I recommend it to anyone.

Eric Lange, Secretary for the NSPKU

The role of Sapropterin (Kuvan®) in the management of PKU in the NHS: considerations and prospects

The advent of dietary treatment for PKU is one of the great successes of modern medicine. Over last 40 years, there has been a dramatic improvement in the outlook for patients with PKU in the U.K.

The core principle of dietary treatment in PKU remains unchanged. However there have been major improvements in several aspects of PKU management such as diagnostics, analytical techniques, food technology, dietetics and psychological and functional assessments. There remains very little research evidence to prove the efficacy of the dietary treatment for PKU (they would be difficult or unethical to do). There is no doubt however, that dietary treatment, despite its drawbacks, has radically improved the outcome in patients with PKU. There continue to be many unresolved issues and unanswered questions regarding the management of PKU.

Adhering to dietary treatment of PKU is not always easy for the patient. Amino Acid supplements are generally not appealing, appetizing or convenient. Dietary restrictions can severely limit the patient's lifestyle. It can lead to significant social and psychological difficulties. It might seem surprising (and disappointing), that after four decades there hasn't been much progress in developing effective and practical alternatives to dietary treatment of PKU. Though there has been continued research with a view to developing alternative modes of treatment for PKU, this has not been translated into tangible benefits for patients.

Even so, there have been some promising recent developments in alternative treatments for PKU. Kuvan® (Sapropterin) is one such mode of treatment, which might have a place in the management of PKU in the U.K.

Kuvan was licensed in the UK in 2009 for the management of PKU and another rare condition known as Biopterin Deficiency. This article specifically discusses the potential role of Kuvan® in the management of PKU.

Phenylalanine Hydroxylase (PAH), the enzyme that is affected in PKU, requires an additional supporting co-factor known as Biopterin for its function. Though patients with PKU do not have a deficiency of Biopterin, in about 20% of patients (about 1 in 5), there was a significant enhancement in the PAH enzyme function when given Biopterin. Therefore, potentially more Phenylalanine (Phe) exchanges could be tolerated in the diet. Biopterin has been used for many years in some countries in patients who have Biopterin responsive PKU.

Kuvan® is a synthetic version of Biopterin and was licensed in 2009.Prior to securing licensing and marketing approval, clinical trials of Kuvan® across several countries were carried out in patients with PKU - children over the age of 4 yrs and in adults. Essentially, these studies showed that

1. A small proportion of patients with PKU showed a significant biochemical response (30% reduction in measured blood Phe concentration) with Kuvan® therapy and that there was a dose-response relationship.
2. There was an increase in the number of protein exchanges that patients could take while maintaining measured blood Phe concentrations or better control of blood Phe.
3. There were no major safety issues in the small number of patients that took part in the trials.

Though it has now been licensed for use in the U.K., it does not mean that patients with PKU can be automatically be treated with Kuvan®. When a new medication is developed and licensed, before it is approved for use within the NHS the following aspects are considered (in no particular order) 1. Cost –effectiveness as compared to existing treatment 2. Cost 3. Efficacy 4. Safety

There are several facts to consider while discussing treatment with Kuvan® -

1. **There is already a highly effective and established treatment for PKU- dietary treatment.** It is a fact that dietary treatment is effective in all patients with PKU without exception. There is no 'failure' of dietary treatment itself. Problems with dietary treatment arise due to difficulties in administration and patient perceptions of the smell, taste and texture of low protein food. Though there has been an improvement in the palatability of dietary products used in the management of PKU, in a small minority of patients there are problems with compliance. There are other problems however which affect the quality of management of PKU- such as variability in the experience of dietetic and medical personnel, motivation of the families, social and factors, ability to cope with dietetic management etc. Therefore the quality of dietary management of PKU is not always satisfactory and is dependent on several factors some of which are complex.

2. **Only a small minority of patients may benefit from** Kuvan®, though almost all patients with PKU show some biochemical response. The definition of 'response' is arbitrary and the reliability of testing protocols is not clear.

3. Kuvan® **is more likely to benefit those with milder forms of PKU** who are already taking a much more relaxed diet compared to those with more severe versions of the condition. The small decrease in Phe in patients with milder forms of PKU may be sufficient enough to remove the need for dietary treatment completely.

4. Kuvan® **is several times more expensive than dietary treatment alone.** This is a major stumbling block in obtaining funding for the use of Kuvan® in the U.K. The cost of Kuvan® based on a dose of 10 mg/kg/day is indicated below. It should be noted that the prescribed dose of Kuvan® could vary between 5-20mg/kg/day. Therefore in some cases the cost could exceed £100,000 per annum per patient. In comparison, the cost of dietary treatment for PKU is on average £10,000 per annum per patient.
   Source -The North East Treatment Advisory Group (NETAG) (NHS) Appraisal Document
   'Sapropterin in the management of Phenylketonuria'
   (Apr 2009)

Body weight (kg)	Actual daily dose (mg) (number of tablets)	Cost of treatment	
		Daily	Annual
15-24	200 (2)	£39.82	£14,534
25-34	300 (3)	£59.73	£21,801
35-44	400 (4)	£79.64	£29,068
45-54	500 (5)	£99.55	£36,336
55-64	600 (6)	£119.46	£43,603
65-74	700 (7)	£139.37	£50,870
75-84	800 (8)	£159.28	£58,137

5. Kuvan® **may not replace the PKU diet** and some patients may need to take a medication and also continue some dietary restriction.

In addition, there are more unanswered questions about the use of Kuvan® in PKU

1. **What is the best definition of a 'response to Biopterin'?** The 30% improvement in Phe concentration is arbitrary and there is no scientific reason behind it. Should anyone with even the slightest biochemical improvement in Phe levels be considered responsive? Or should response only mean that Kuvan® has taken away the need for dietary management completely in a patient?

2. **What is the best method to test for Biopterin response?** Should the response be tested after a single dose, one day or several days? How do we know if patients are consciously or subconsciously making alterations to their diet while being tested (which might affect their Phe concentrations)? Studies show that the results of various testing methods described in the literature are poorly comparable and not always reproducible.

3. **How long should treatment with** Kuvan® **be continued?** Should patients be treated with Kuvan® until the age of 12 yrs (there is currently no evidence of an adverse effect on I.Q if PKU treatment is discontinued after this age)? At 16 years (beyond the paediatric age-group)? After higher education? Should it be given for a short time to achieve specific goals (such as exams) so that they can concentrate less on their diet?

4. **Is Kuvan® safe and effective in infants and children under the age of 4 yrs?** Kuvan®, at present, is not licensed for use in children under the age of 4yrs. The quality of management of PKU in the early years is probably more crucial than during later years. Safety data has only been recorded in small numbers of children over a short period. There was a decrease in White Blood Cell counts (neutropenia) in 4 % patients on treatment with Kuvan® during the clinical trials. Interestingly, Sapropterin (Kuvan®) is also being currently considered for other indications such as in the treatment of high blood pressure in adults. It is not clear whether Kuvan® has an unintended effect on blood pressure in patients, especially children, with PKU. Clinical trials are currently being planned to establish safety and efficacy in children under the age of four.

5. **Is** Kuvan® **safe for use in Pregnancy?** Kuvan® has been used in pregnancy when the mother's Phe control has been poor in order to protect the unborn child. The benefits and risks of Kuvan® therapy in pregnancy have not been established.

6. **Does** Kuvan® **lead to a measurable improvement in the quality of life for patients with PKU?** Parents and patients may see a remarkable improvement from Kuvan® treatment in their outlook, mood, life, performance, energy levels, relationships, appetite etc. However this may be difficult to quantify. Separating scientific fact from emotion, human nature and placebo effect is not easy. The benefits of Kuvan® therapy to patients' quality of life are largely anecdotal and subjective. Clinical trials have been too short to provide any significant long-term data regarding 'real-world' clinical efficacy. Further studies are required to establish and quantify these, often, intangible benefits. Long-term studies into these aspects of treatment with Kuvan® are under consideration.

7. **The long-term safety of relaxation of the diet in these patients is not known.** The intention of the clinical trials was not to assess the ability of Kuvan® to enable liberalization of the PKU diet but to study the ability of Kuvan® to improve the management of blood Phe concentration in patients with poor control. Some may see relaxation of the diet as a promising benefit of Kuvan® therapy, but this cannot be recommended without caution. There is no evidence for the safety of dietary relaxation or other dietary changes in patients with PKU treated with Kuvan®.

There are still several 'unknowns' regarding the role of Kuvan® in the management of PKU. It has been, therefore, very difficult even for professionals to understand and agree regarding the potential role in the treatment of PKU or Kuvan® in the U.K. It is a telling fact that even in countries where Kuvan® is currently used in the treatment of PKU, there are no published national or regional guidelines for its use (in many countries there is no guideline for dietary management). In most centres, treatment decisions are driven almost entirely by the patient's choice and the doctor's discretion. The population characteristics of patients with PKU, the scrutiny that these decisions receive, the level of evidence required to support treatment decisions, the nature of the healthcare systems and methods of funding in those countries are different from those in the U.K.

Ideally, metabolic teams across the U.K would like to be able to test all eligible patients with PKU for a response to Biopterin and treat those who respond according to agreed criteria. There is no doubt regarding the short-term biochemical efficacy of Kuvan® in a small minority of patients. Long-term studies are required to establish clinical effectiveness. Assessment of cost effectiveness of Kuvan® however, requires the expertise of health economists and commissioners. The decision whether rare treatments such as Kuvan® should be approved for funding is not easy. The NHS, the Government and the wider society should carefully consider the relevant financial, ethical and moral issues in addition to current medical evidence and knowledge.

The budget of the health service is very similar to a family's finances. There is a finite amount of money and a long wish list of priorities. The health service therefore looks for value for money when new services or medications are funded and seeks to invest in areas where there is likely to be the greatest measurable health benefit in the community.

At present there are no nationally agreed guidelines for the use of Kuvan®. Efforts are on to produce consensus guidelines for the use of Kuvan® in specific groups of patients with PKU. It is hoped that such guidelines will assist commissioners and therefore the NHS in decision-making and facilitate access to Kuvan® for those deserving patients.

Until these guidelines are accepted and a mechanism for funding Kuvan® therapy for specific indications is established, the only option currently available to patients and metabolic teams is to make an application for Exceptional Case Funding (ECF) of Kuvan® therapy to Healthcare Commissioners. Evidence will be required to show that the indication for treating the patient with Kuvan® is truly 'exceptional' and it is not a new service development generally for any group of patients with PKU.

The NHS in Wales and Scotland does not recommend Kuvan® for use in the treatment of PKU as the marketing authorization holder (Merck-Serono) did not possess enough evidence/data to make a submission. In England, NICE (the National Institute for Clinical Excellence) did not consider Kuvan® suitable for appraisal on similar grounds. The rarity of PKU (especially Biopterin-responsive PKU) makes it difficult to assess clinical and cost-effectiveness of Kuvan® in the same way as other more common treatments. Kuvan® is an 'Orphan Drug'. The Orphan Drug law seeks to encourage development of new therapies for rare medical conditions, which would otherwise be unprofitable to develop. Pharmaceutical companies that develop such treatments are provided tax-breaks and a monopoly for 7 to 10 years. The enormous cost of development of Orphan Drugs for a small group of patients means that these new drugs will invariably be very expensive.

The prospects for funding of Kuvan® in the U.K are likely to improve if further evidence becomes available regarding clinical effectiveness and if treatment with Kuvan® becomes a cost-effective alternative to dietary treatment. In the future, it is likely that there will be greater scrutiny, stricter guidelines and increasing pressure to ensure economy and cost-effectiveness before new, more expensive medications are approved for use within the NHS.

Continued support from clinical teams, sustained lobbying by patients and the NSPKU will be crucial in ensuring that patients with PKU in the U.K. benefit from new developments in treatment.

By Dr Suresh Vijay, Consultant in Metabolic Disorders, Birmingham Children's Hospital

Healthy Living on a Low Protein Diet

There are so many healthy eating messages out there, but how do you apply it to a low protein diet? This is the question we tried to answer at our healthy living event in Birmingham on a Saturday afternoon!

We decided to hold the event after we carried out an audit in the UK to look at the body mass index of people with PKU who follow a low protein diet. We looked 269 people from centres in Northern Ireland, Birmingham, Manchester, Sheffield, Cambridge, Glasgow and London. We found that people with PKU following a low protein diet were just as likely to be overweight as the general population. This is quite worrying as over 50% of the UK population are overweight. Being overweight can increase your chance of developing heart disease and diabetes and your diet could become even more complicated.

At the healthy living event we welcomed everyone with fruit juices, smoothies, fruit kebabs and veggie dips and then everyone listened to a talk on healthy eating. The talk included tips on how to work out what foods were high in fat and sugar and what foods to choose instead.

This was followed by 2 workshops, a low protein healthy eating cooking demonstration and an exercise class.

PKU low protein food pyramid

Left to right: Jane Lodwig, Specialist Nurse, Louise Robertson, Specialist Dietitian and Dr Tarek Hiwot, Consultant in Inherited Metabolic Disorders at University Hospital Birmingham NHS Foundation Trust

Mona doing her cooking demonstration.

Ewan showing us how to do our exercises!

Mona Taylor, Home Economist for SHS Nutricia, did a brilliant cooking demonstration. She showed us how to make filled tomatoes, risotto, Thai green curry soup and fruit smoothies. She also gave us valuable low protein cooking tips!

Ewan Forbes, from SHS Nutricia, gave us a gruelling 30min circuit class. He taught everyone how to do exercises with resistance bands. Everyone was lucky to be given a resistance band to take away with them to practice their exercises at home!

Everyone enjoyed themselves and hopefully learnt a thing or two as well.

A big thank you to Tina Aniyi and her team from SHS Nutricia who helped organise and sponsor the afternoon.

If you would like to lose weight, then try these tips to decrease the calories in the diet

- Choose low fat products
- Cut down on the amount of oil you use in cooking
- Use pickles or chutneys in sandwiches instead of margarine or butter
- Choose diet drinks that have the sweetener sucralose instead of aspartame (check out supermarket own brand drinks)
- Cut down on sugar in hot drinks, try a sweetener (without aspartame)
- Choose fruit or dried fruit as snacks instead of biscuits and crisps

Have a go at reading the fat content on food labels to see if the foods you are eating are high or low fat.

Label reading

High fat foods	More than 20g of fat per 100g of food
Low fat foods	3g of fat or less per 100g of food

Louise Robertson,
Specialist Dietitian in Inherited Metabolic Disorders,
University Hospital Birmingham NHS Foundation Trust.


Loprofin 4 Part Baked Bread Rolls
Loprofin Crunch Bar
8
Loprofin Mix
Loprofin
SNO-PRO


Loprofin Fusilli
500g
Loprofin Rice
500g
Loprofin Loops
Loprofin Animal Pasta
500g
Loprofin Dessert
150g
Loprofin Cake Mix
500g


Loprofin Wafers
Loprofin Wafers
100g


NUTRICIA
SHS-Homeward

Festive Recipes with...

Loprofin

Christmas Biscuits

Ingredients

(makes approximately 20 biscuits)

½ Loprofin Low Protein Duo Bar, flavoured

50g (2oz) butter

40g (1½oz) hard margarine

50g (2oz) caster sugar

175g (7oz) Loprofin Low Protein Mix

4 x 5ml (4tsp) water, approximately

Oven Temperature: 150°C/300°F/Gas Mark 2

Method

1. Grate half the Loprofin Low Protein Duo Bar

2. Place the butter, margarine, and grated Duo Bar into a mixing bowl and beat well; add the sugar and beat together until light in colour and texture

3. Stir in the Loprofin Low Protein Mix, using a wooden spoon

4. Using one hand, squeeze the mixture together until it begins to bind, adding sufficient water to give a manageable dough; transfer to a surface lightly dusted with Loprofin Low Protein Mix and knead the dough for about 30 seconds, until smooth

5. On a surface lightly dusted with Loprofin Low Protein Mix roll out the dough to about ½ cm (¼ inch) thickness

6. Using a selection of Christmas biscuit cutters, cut out shapes from the dough; lightly knead, re-roll and cut out remaining dough

7. Transfer the biscuits to lightly greased baking trays and bake in a preheated oven for 20-25 minutes, until pale golden in colour

8. Remove from the baking trays whilst still warm, cool on a wire rack

9. When the biscuits are cold, decorate as desired - using either melted Duo Bars or coloured water icing and colour sugar sprinkles*

Note

To harden the Duo Bar quickly place the biscuits in a freezer for a few minutes

* check NSPKU food list for suitable brand

Christmas Pudding

Ingredients

(makes 6-8 portions)

50g (2oz) Loprofin Low Protein Mix

25g (1oz) breadcrumbs from a Loprofin Low Protein Loaf

65g (1½oz) dark muscovado sugar

65g (1½oz) vegetable suet

1 x 5ml (1tsp) ground mixed spice

1 x 1.25ml (¼tsp) bicarbonate of soda

1 x 1.25ml (¼tsp) salt

1 x 5ml (1tsp) orange rind, finely grated

1 x 5ml (1tsp) lemon rind, finely grated

200g (8oz) mixed dried fruit

100g (4oz) baking apple, peeled, cored and coarsely grated

1 x 15ml (1tbsp) black treacle

3 x 15ml (3tbsp) Loprofin PKU Milk Drink, or spirit of your choice

½ litre (1 pint) pudding basin

Method

1. Place all the dry ingredients, including grated fruit peel, mixed dried fruit and apple in a large bowl and toss well together

2. Mix together the black treacle and Loprofin PKU Drink (or spirit if preferred)

3. Stir the treacle and Loprofin PKU Drink into the ingredients in the bowl, beat for a few seconds with a wooden spoon, until thoroughly combined

4. Transfer the mixture to a ½ litre (1 pint) pudding basin, press well down and level the top, using the back of a wooden spoon; cover the basin securely with a lid, greaseproof paper or foil

5. Steam the pudding for two hours, then serve with a Low Protein white sauce or custard

Note

To reheat individual portions of pudding, cover and microwave on full power for 1-1½ minutes (depending on the wattage of the microwave)

Steaming

This is usually done in a 'steamer' pan - a pan comprising of two sections; the steamer part has holes over its base, allowing steam from the water in the pan to come into contact with the basin and cook the pudding. However, if you do not have a steamer, fill a large saucepan with sufficient water to come halfway up the basin, put an old saucer or crossed skewers on the base of the pan to keep the basin off the bottom. During steaming keep the water gently bubbling and replenish with boiling water as it boils away

Ed's Recipes

Creamy tomato soup

INGREDIENTS

- 1 onion, chopped
- 25g butter
- 1 clove garlic, crushed
- 1tsp paprika
- 1tbsp tomato purée
- 1kg tomatoes, chopped
- 2tsp sugar
- 1 bay leaf
- 600ml vegetable stock
- 142ml pro zero
- Basil leaves, to serve (optional)

METHOD

Fry the onion in the butter for 10 mins until softened. Stir in the garlic and paprika and cook for 1 minute. Add the purée and continue for 2 mins more.

Add the tomatoes, sugar, bay leaf and vegetable stock. Bring the soup to the boil then simmer for 20 mins.

Remove the bay leaf. Blend the soup in batches in a food processor or blender until smooth. Push through a sieve to remove any skin or seeds and return the liquid to the rinsed-out pan.

Stir in two-thirds of the sno-pro and reheat the soup gently. Once it's hot, serve your tomato soup immediately in warmed bowls with a swirl of the remaining sno-pro and a garnish of basil leaves.

Carrot and honey soup

INGREDIENTS

- 2 tbsp butter
- 2 small leeks , sliced
- 800g carrots , roughly chopped
- 2 tsp clear honey
- small pinch dried chilli flakes (optional)
- 1 bay leaf
- 2½ l vegetable stock

METHOD

Melt the butter in a large saucepan over a medium heat. Add the leeks to the pan, then cook for 3 mins until starting to soften. Add the carrots, honey, chilli (if using) and bay leaf, then cook for 2 mins.

Pour in the stock, bring to the boil, then simmer for 30 mins. Blend the soup in batches, return to a clean pan, then season to taste. When ready to serve, bring back to a simmer, then ladle into mugs

Dietitian's Report

Chocolate Flavour Surprises

Chocolate, as we all know, is very more-ish and has a high protein content. It's one of those foods which we advise to avoid because keeping to 10g of chocolate for an exchange is very difficult and once tried it is very hard to accept the low protein equivalent.

However, recently two mothers of children with PKU have found some very surprising and potentially useful chocolate **flavoured** foods in the supermarket.

Barbara McGovern told me about some little white sweet drops which are covered in hundreds and thousands from Tescos. They are called White Snowies and have the appearance and texture of white chocolate but are not chocolate. They contain 0.3g protein per 100g. This means that they do not have to be counted as an exchange. The bags contain 70g and is quite a lot – half a bag would be a good helping. They are found in the '3 bags for £1' section amongst the sweets for children. The protein content has been confirmed by Tescos.

Louise Brennand also found something useful- a white chocolate flavour cake covering by Silver Spoon for melting over cakes and biscuits. She was quite rightly surprised to find that the protein on the pack indicated that it was 0.6g protein for 100g. (Real chocolate has a protein content of 7-8g per 100g). However, this is a chocolate flavoured cake covering which does make the difference. Foods can be called 'flavoured' but not have to contain the real thing e.g vanilla flavoured ice cream does not have to contain real vanilla.

Silver Spoon have confirmed in writing that the protein content of 0.6g per 100g is correct and the suppliers have informed Silver Spoon that it is low because they use de-proteinised whey powder.

However, Silver Spoon have informed me that they are sourcing the white cake covering from 2 suppliers as they have had a large demand for it. The second supplier's white cake covering has a higher protein content of 3g per 100g, they are clearly labelled with the protein content and are much shorter and fatter. It is important that you check the protein content on the back of the bars.

White Chocolate Flavour Cake Covering Protein 0.6g per 100g

1 exchange = 165g

White Chocolate Flavour Cake Covering Protein 3g per 100g

1 exchange = 35g

Betty Crocker Chocolate Flavour Buttercream and Chocolate Fudge Flavour Icing

These two cake icings had a protein content of:

Buttercream Style Chocolate 0.46g per 100g

Rich and Creamy Chocolate Fudge 2.2g per 100g

The recipe has changed and the nutrition labels now state 0g protein per 100g. I have asked the company to confirm in writing that this is correct but so far have not received this and there appears to be some confusion about protein content. We have decided to have the 2 icings analysed for phenylalanine content so that we know for sure what the level is. In the meantime you can use the Buttercream Vanilla and Rich and Creamy Vanilla without counting as an exchange.

Goody Good Stuff

Barbara Cochrane, dietitian in Glasgow, went to a food fare recently and came across a company called Goody Good Stuff which makes vegetarian sweets which are –Fat free, meat free, dairy free, gluten free, egg free, nut free, soy free, gelatine free, kosher, Halal and no artificial colours and flavours –what a list of 'frees!' Anyway they are also protein free. They are gummy sweets which come in 8 flavours and can be found in Health Food shops. They are not calorie free or sugar free – sadly for some of us!

Nestle Curiously Cinnamon Cereal

Louise Brennand also told me about a new breakfast cereal which is wholewheat and which her daughter loves. The protein content is quite low at 4.9g per 100g. So that's 20g=1 exchange.

She said her daughter really liked the different flavour. I bought a box of them and my own daughter (non-PKU) had to remove them from sight she liked them dry from the packet so much. Breakfast is such an important meal and if you struggle to get your child to eat some cereal you might like to try this one.

Amino Acid Analysis

We would like to get some more foods analysed for phenylalanine content and I would like your suggestions. It is better to have un-processed foods as manufactured foods are sometimes only on the shelves for a short time. Is there anything you have come across which you've wondered about? Let me know and I can put it on the list of suggestions. It's always nice to try new things and I have encouraged my family to do so but my youngest one (she doesn't have PKU) has just come back from travelling and I was in full agreement with her over reluctance to try a fried mouse. There are limits.

A short break in Skipton

In August, my husband and I took our grandson Jack (14, PKU) and nephew Rory (16) to spend a few days in the historic market town of Skipton, North Yorkshire. This is a beautiful part of the country, and has a lot to commend it for a holiday.

We chose the Rendezvous Hotel, about mile outside the town centre. The NSPKU has held its annual conference there several times, so we knew that the hotel chef understood the PKU diet. On booking we found it reassuring that even the receptionists knew about PKU, and appreciated that we would need special food.

I contacted David, the head chef, about a week before we went, and we discussed the dishes he might make for Jack, and which items I needed to take. David said he could make "anything you want, really", which was delightful to hear. We'd booked two nights at the hotel, so we planned two dinner menus. I gave the staff some lasagne sheets, a packet of Fate all-purpose mix and a large carton of Prozero milk replacement, together with some photocopied sheets reminding them which foods were, and were not, allowed. I baked a dozen bread rolls the day before we went, and six of these were put in the hotel fridge and six in the freezer as soon as we arrived.

The chef and I had not really discussed breakfast (as it's not a usually a problem), but David asked me if I had brought any sausage mix which he could prepare for Jack's breakfast. Sadly, I had had not: I make plenty of Fate sausages at home but, to be honest, I didn't know that sausage *mix* existed. It would have been useful, and I'll remember it for next time! In any case, the hotel offers a comprehensive breakfast buffet, so Jack had an exchange of cereal or hash browns, grilled tomato and mushrooms, plenty of fruit, and the usual toast and preserves.

I made a picnic lunch each day (salads, bread, honey, fruit, crisps, and home-made cake and shortbread) which bridged the gap between breakfast and dinner.

In the evenings, David cooked some lovely things for Jack, including a 3-melon starter, an avocado salad, marinated kebabs in a delicious tomato and chili sauce, mushroom lasagne, apple crumble, and a sorbet and baked fruit dessert. Jack really enjoyed his meals. The rest of the family ate well, too, and it was lovely for us all to be able to sit down in a restaurant and not worry about Jack's food.

There are lots of things to do around Skipton, a town known as "the gateway to the Dales". Skipton Castle, over 900 years old and one of the best preserved medieval castles, stands in the middle of town and is a great tourist attraction. We enjoyed a canal boat trip on the Leeds-Liverpool canal in glorious sunshine; took a trip on the Embsay to Bolton Abbey steam railway; and explored the beautiful River Wharfe around Bolton Abbey. We watched herons there, and marvelled at the force of water at "the Strid", the point at which a huge volume of water is pressured through a very narrow channel. The rocks have been hollowed out here and shaped into strange forms by the dynamism of the river.

The hotel swimming pool was a tremendous attraction for the boys (and would have been a godsend if the weather had been bad). They swam twice a day, and really enjoyed it. All in all, we had a delightful little holiday at the Rendezvous in Skipton.

Barbara Broadbent

Congratulations

and thank you to all our competition winners!

NEW design for 2010

Laughlan, age 4, Glasgow	**Benjamin,** age 7, Liverpool
Amy, age 8 Manchester	**Carl,** age 9, Liverpool
Isla, age 6, Glasgow	**Nadia,** age 9 ¾ , Newcastle upon Tyne
Sophie, age 6, Isle of Mull	**Scott,** age 6, Glasgow
Chloe, age 12, Doncaster	**Molly,** age 4, Birmingham
Adeline, age 2, La Roche, Belgium	**Caria,** age 6, Bradford
Abigail, age 4	**Sophie,** age 8, Quebec, Canada
Iain, age 8, Glasgow	**Annabelle,** age 10, Bradford
Callum, age 5, West Midlands	**Rebeccah,** age 8, Glasgow

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Quick Fudge

Equipment needed

Large bowl
Saucepan

Ingredients

100g chopped **Vitabite**™
50g butter
30ml **PROZERO**™
5ml vanilla essence
425g icing sugar

Instructions

1. Melt the Vitabite™ and butter in a bowl over a pan of hot water or alternatively in a microwave.
2. Stir in the **PROZERO**™ and vanilla essence.
3. Gradually stir in the icing sugar.
4. Put into a lightly greased square baking tray and spread evenly.
5. Allow the fudge to set in the fridge then cut into bite-sized chunks.

Serving suggestions:

Why not decorate the fudge with permitted Christmas theme icing decorations? Or add some dried fruit when making up for fruity fudge!

Other great ideas for Christmas with VITABITE™

Christmas tree decorations

Pour melted **Vitabite**™ into a chocolate mould. Allow to set, then wrap in coloured foil so you can hang them on your tree!

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NSPKU 38th Annual Family Weekend & Conference 2011

The Britannia Adelphi Hotel Liverpool

Friday 18th March to Sunday 20th March 2011

Venue:

We have a new venue for this year's conference, the historic Adelphi Hotel in Liverpool.

The Adelphi Hotel is beautifully located in the heart of Liverpool in the North West of England. This internationally renowned hotel is luxurious, well appointed and a superb base from which to explore the many tourist and business attractions of Merseyside. Built in 1914 with superb public rooms and panelled suites, the Britannia Adelphi is Liverpool's largest hotel. It was recently refurbished and now contains all of the modern facilities and amenities that today's traveller requires yet still retains its historical appearance and significance.

Accommodation:

The hotel has a wide selection of bedrooms, meeting rooms and lounge areas.

Supervision of Children:

Whilst the children will be supervised in the crèche, the kids' club and outings, it is not possible for the Society or hotel staff to supervise or be in any way responsible for the children at other times or places. It is the responsibility of parents to supervise their children.

Facilities:

The Britannia Adelphi boasts stunningly decorated and furnished guest bedrooms, all of which are en suite. Each room contains, as standard, tea/coffee making facilities, hairdryer, trouser press and satellite television. Additionally, the larger rooms and lounge areas within the hotel offer high speed broadband internet access.

PKU Diet:

You will need to bring your own protein substitutes, vitamins and minerals. The diet will otherwise be catered for. A limited number of baby foods will be available. If however, your infant prefers certain foods we would ask you to bring these along with you. The PKU diet will be catered for from Friday evening to Sunday lunchtime inclusively. It will not be available for any additional nights.

Location:

Air: Liverpool John Lennon Airport is in easy reach of the city centre by train

Rail: Liverpool Lime Street just 2 minutes walk.

Road: Parking is limited and charged extra.

Assisted Places:

There is restricted funding available for those who require financial assistance with conference fees. Please enquire in the first instance to the conference organiser who will help you with your enquiry. All applications will be dealt with in the strictest confidence.

Family Weekend / Conference Fees:

Non-PKU Adult (17 yrs and over)	£210.00
PKU Adult (17 yrs and over)	£105.00
Non PKU Child (5 - 16)	£105.00
Non PKU Child (0 - 4)	£50.00
PKU Child (0 - 17)	Free
All under 4's	Free

Day Visitor Fees (including all meals):

Saturday	£40.00
Sunday	£20.00
PKU Adult (17 yrs and over)	Half above rate
PKU Child (0 - 17)	Free

Additional Nights:

If you wish to stay an extra night before or after the conference, please contact the hotel & book direct. We have negotiated a special rate for Bed and Breakfast of £50.00 per single room, £80 per double (£10.00 per child under the age of 15 years if sharing above). This rate is for Thursday or Sunday night.

The NSPKU Conference Organiser is David Stening **chair@nspku.org** Tel 0207 099 7431 www.nspku.org

All PKFoods are now available on prescription

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To know that we really understand PKU, really understand your diet and really know what exchanges are?

Yes... we do!

Working closely with PKU dietitians and PKU patients Leanora Munn and Pauline Young of Yes We Cook That.com now have a menu of 180 ready meals and snacks all suitable for PKU diets.

All meals are clearly labelled with the ingredients and any exchanges and are made using only prescriptive flours, egg replacers, milks and pastas.

Our menus are available through your dietician in some parts of the country or directly from Yes We Cook That.com

Our aim is to supply all PKU people with the opportunity to purchase ready made meals, breads, cakes and sweet and savoury snacks at supermarket prices.

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