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**The Council of Management**

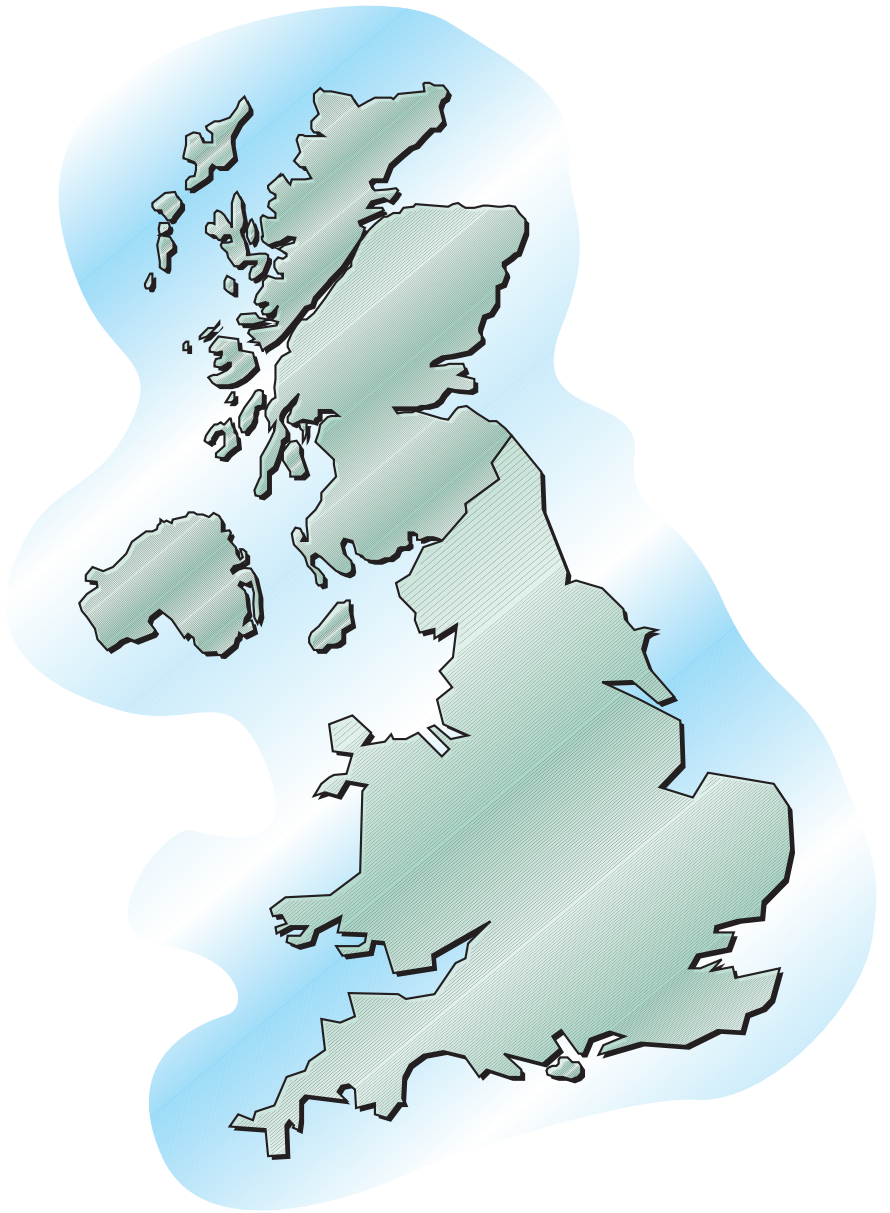
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## Editor's Report...

Dear Readers,

I thought I should introduce myself and family to you as I have, with some trepidation, agreed to take over the role of Editor of News and Views from Kiri Thomas. She has been absolutely brilliant over the last 5 years and I only hope I can live up to her reputation.

I have a 19 year old daughter, Kylie, who was diagnosed with PKU at the age of 3½ in South Africa. You can only imagine the trauma that was. However after phoning Great Ormond Street Hospital, the staff there put us in touch with the NSPKU and we have never looked back. We received our magazine faithfully every quarter and were so inspired by recipes and all the articles written that we chose to ignore her prognosis and "get on with it!" We decided to move to England in 1999, when she was 10 years old, as our limited supply of low protein foods was becoming increasingly difficult to obtain making her diet impossible to maintain in South Africa.

Since coming here, we have made many friends through the NSPKU and Kylie has thrived becoming a confident and happy young adult who has defied the text books. She has successfully completed 2 college courses and will soon be seeking full time employment in the Animal Care industry.

We are extremely grateful to the NSPKU for its unending support over the years – we know she would not be where she is today without it.

My aim as editor, apart from bringing you as many interesting articles etc as possible, will be to add recipes from the last 16 years of low protein cookery. I have always tried to make Kylie's food look like ours and will include recipes in future issues.

I look forward to working with the COM on future issues and hope I can do them proud.

**Anne Verster,**  
Editor



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Layout by **The Keystroke Mill**  
[www.keystrokemill.com](http://www.keystrokemill.com)  
Tel: 01444 236035

Printed by action  
Tel: 01444 236204



# Letters

News & Views, 23 Barnfield Cottages, Arundel Road, Fontwell, West Sussex BN18 0SE  
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*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,

As I suffer from PKU, my friends and I organised a charity five a side football tournament at our school, Long Ditton St Mary's Junior School, to raise money for NSPKU.

I am pleased to enclose a cheque for £205.76.

I also enclose a photograph of me and my fellow house captains, Jessica Wenlock and Luke Mullaney celebrating the event at the finish.

Yours sincerely  
Beth Garner (aged 10)



Dear News and Views,

Our grandson George, 4 months, has PKU so we thought we would try some fundraising and held a "Good as New" book sale. We had over 400 books to sell along with tea, coffee and cakes. 40 people called in and £400 was raised. A satisfactory day. As you can see after 6 hours George was feeling slightly bored! Photo attached.

Yours,  
Lyn Houghton



Dear News and Views,

Since our beautiful daughter Rebekah was diagnosed with PKU last October we have been extremely grateful for the support and information we have received from NSPKU and its partners and our dietitian team. We know this is just the start of a journey that won't be without some difficult moments, but with this continued support we are feeling up for the challenge.

To show our gratitude we are proud to be donating £1871 to NSPKU. This fantastic amount has been raised through two events; firstly Rebekah's Auntie Kimmy and Auntie Amy completed the Bournemouth half marathon in April, both impressively beating 2 hours, and laying the gauntlet down to dad now!

Secondly at the end of April we had Rebekah christened at St John's in Keynsham and requested donations to NSPKU rather than gifts. We have been overwhelmed by the generosity of our family and friends.

Keep up the good work.

Andy, Helen and Rebekah Muncer



Dear News and Views,

Would you please pass on the following email address to anyone who would be interested in PKU contacts.

World Link contacts email at [www.PKUworldlink.org](http://www.PKUworldlink.org) .

There doesn't appear to be any Northern Ireland contacts and if anyone would like to contact me, please email me at my address at the top of this letter.

Thank you, Rory O' Honlon



 Dear News and Views,

Please find enclosed a cheque £120.00 this is the amount that has been donated to the NSPKU from my family and work colleagues.

I have just attended the conference in Torquay with my daughter Alicia who is 7 years old and has PKU. This was the 1<sup>st</sup> conference that we have stayed at, last year we went as day visitors as it was held in our hometown of Northampton. I would just like to say a big Thank-you to all of the people involved in organizing this event.

We had such a wonderful time. It was a pleasure to see Alicia making friends with children who are the same as her, making her realize she is not the only person in the world with this challenging condition.

She thoroughly enjoyed the day trips that she went on and on our return journey announced that she didn't want to go home because she was having such a good time. For me the benefits were I didn't have to worry about her food-as this was all catered for, I had a happy little girl and we had a weekend of fun. For anyone who is wondering whether to attend a conference in the future, I can honestly say in the two times that we have attended I have learnt such a lot in such a short time, have met some lovely people and been offered great support, my only regret is that we didn't attend one before now, as we have struggled at times with Alicia's compliance and have not really had much voluntary information and support from our area. I am looking forward to the next one.

**Yours sincerely**  
**A.M. Jeyes**

 Dear News and Views,

Since this Company formed in March 2000, each year at Christmas donations are collected from staff in lieu of sending one another Christmas cards. The staff are also requested to nominate their preferred Charity to benefit and have an option to suggest alternatives. One of our members of staff suggested your charity and, following a ballot, it resulted in being one of the charities to benefit this year.

Additionally, we hold a staff raffle of items gifted to the Company over the festive period the proceeds from which is added to this fund.

As a result of these initiatives, I am now pleased to enclose a cheque for £350 as part of the proceeds from this fundraising.

Yours sincerely,

**Ray Brookes**  
**Group Chief Executive, The Community Housing Group**

## “How many times have you been to a wedding and come across five PKU guests?”

On a sunny day in May 2008 friends and family gathered together for Andy Cardy and Heather Paine's wedding at St John's Church in Felixstowe. Among those attending were five people with PKU (two of whom were the groom and best man!) The wedding itself went really well and afterwards the guests went to a reception nearby. The PKUs were well catered for at the dinner, enjoying vegetable soup followed by a roast with stuffed mushrooms, then fruit salad or a “Cheezly” cheese board.

In the evening the buffet had a PKU section of its own, with PKU pasta salad, a couscous dish, and other tasty items. It was enjoyed by all.

Thanks Andy and Hev for the PKU hospitality and we wish you many happy years together.

Happy Anniversary from Paul & Julie



find  
us  
on  
the  
web  
at:

www.nspku.org

## Detailed Statement of Financial Activities for the Year Ended 31st October 2008

	31.10.08 £	31.10.07 £
<b>INCOMING RESOURCES</b>		
Donations & sponsorship	49,893	65,556
Sundry income	240	-
Fundraising events	4,145	7,196
Advertising	5,100	9,450
Deposit account interest	6,725	4,786
Membership & literary fees	9,294	10,578
Income from conference & events	3,772	5,298
Publications income	1,492	4,217
Exceptional items	6,500	-
<b>Total incoming resources</b>	<b>87,161</b>	<b>107,081</b>
<b>RESOURCES EXPENDED</b>		
Merchandising & fund raising costs	1,342	850
Conference & event costs	17,458	14,224
Newsletter & publications	20,886	19,302
Research	2,901	2,681
Advertising	-	108
Sponsorship	706	705
Assisted places	1,100	576
Accountancy	1,025	705
Auditors' remuneration	1,025	705
Dieticians fees & expenses	14,604	15,330
Insurance	833	1,847
Administration fees & expenses	7,053	5,304
Meeting & conference expenses	6,236	6,609
Membership fees & subscriptions	461	523
Telephone	1,366	1,092
Travel	3,064	4,114
Postage & stationery	1,497	1,930
Depreciation of tangible fixed assets	197	319
<b>Total resources expended</b>	<b>81,754</b>	<b>76,924</b>
<b>Net income/(expenditure)</b>	<b>5,407</b>	<b>30,157</b>

## Yes We Cook That.com

Specialist Dietary Foods

# Freshly prepared PKU ready meals

Now available by post to all UK and Northern Ireland addresses

### Would you like...

To open the fridge to a freshly cooked PKU meal, labelled with the ingredients and exchanges waiting for you?

### Yes We Cook That!

To open the freezer and find Spaghetti 'Meat' balls and Fishless Fingers and Chickless Nuggets, all with 0 exchanges?

### Yes We Cook That!

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 dietitian 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.

**Phone Leanora or Pauline on 07773 342594**

24 hour answering machine 01290 332 949

e-mail [sales@yeswecookthat.com](mailto:sales@yeswecookthat.com)

Yes We Cook That.com, 39 Leggate, New Cumnock, Ayrshire, KA18 4NG

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# A letter from Cape Town

Hello everyone!

I'm currently 9 months of the way through my year of voluntary work in a Children's Home in South Africa, and thought I would let you know how my work and diet are going.

My volunteer placement was organized by Project Trust, a charity organization based on the Isle of Coll, off the north coast of Scotland. Following a presentation from the charity representative at my Sixth Form, I knew it was something I really wanted to do before continuing with further study. I then went for a week to the island for selection and once I was accepted I spent another week there to have essential training.

There were 2 big issues to consider before I could go. The fundraising seemed daunting – almost £5000 to be raised. We started with a sponsored swim – dad got all his friends involved! Then continued with a Quiz night, a Glam Rock night... somehow the money totalled up with the very generous donations from family and friends in England and Ireland.

Next my food! The Director of Project Trust was concerned about how I would manage with my diet and what would happen if I could not obtain the appropriate food. After a number of phone calls and with the help of my dietitian and consultant at the Royal London Hospital, the Trust was satisfied that I would manage. It never really worried me - after having been to the annual PKU conference every year, I'd heard all about the experiences of other PKU travellers and knew my diet would not be a problem.

We approached SHS at the PKU conference in 2008 and they agreed to not only send me my food supplements and low protein food but also to give me very generous sponsorship. There were a few hiccups with my supplements at Customs in South Africa as they refused to release my food for a few weeks. The SHS representative was excellent and contacted me and my parents regularly to reassure us

that he was sorting it out and advised us not to pay any money to release the food to customs directly if we were contacted – apparently they have been known to do this! I had been out of supplements for just a couple of days when they did reach me so it all went well. I take my food supplements daily and rely heavily on low protein pasta – not only is it good for my levels but it is also very cheap when you are on a very tight budget! I eat a lot of stir fries as well but I don't think I will ever eat butternut squash in any form again when I go home – it comes mashed, fried, baked, boiled, roasted, steamed, with sugar, cinnamon or salt...and it's on the menu every single day!

I've been working almost non stop in the Children's Home since I arrived on the 1st September 2008. It is a suburban Children's Home housing 144 children from the age of 2-19. Most of the children are not actually orphans but have had to be removed from their family





homes because they were unsafe. Two thirds of them have suffered from sexual abuse while many were taken off the street and have Alcohol Foetal Syndrome. This is very sad and sometimes hard to deal with as it obviously has a great impact on the children's mentality and behaviour.

We have 12 houses on the grounds and they are divided by gender but mixed in age to create a family atmosphere and keep siblings together. Last year all the international volunteers (20 of us) lived together in one of the houses and that was an experience of its own as well as entertaining the visiting cockroaches and rats! The majority of the volunteers here are German but there are also some from Holland, Panama, Brazil and Scotland. Most of the volunteers know about my diet. It's not something I'm shy about, although I don't feel there's a need to make a big deal out of it. Everyone was interested, but never rude although I do get a lot of jokes about my 'fake food'.

Since early this year the home has been restructured because sadly some of the children are seriously out of control, and the police have had to be called here many times. Two houses have been converted into a secure care unit housing 7 boys and 7 girls who have severe mood or behaviour disorders. It's not a prison but it is surrounded by high walls and electric fences as sometimes the children are violent and attempt to run away. They have a very structured day and a detailed programme with a permanent social worker. Animal therapy has been introduced to help them - it is believed that if they can learn to give and receive love from animals they will later be able to do so with people. These children have been through so much that they often lack the ability to communicate and deal with their emotions and have difficulty with the concept of right and wrong.

This year, I am living in a house just with children aged 6-18 years. I share the kitchen and bathroom with them but have my own room. I have got to know them so well that they feel like my own little family! They are also very interested in my diet, I've explained it to them as simply as I can and they have lots of fun tasting my food, but they weren't so impressed with my supplement!

As a volunteer I work where I am needed most and do both day and night shifts. In the evenings I help with homework, read to and play games with the children and put them to bed. In the morning we get the children ready and take them to their schools. During the school day I look after the very young children as well as those that cannot find a place in school or were expelled due to their aggressive behaviour.

As volunteers we are lucky to get involved with outreach projects with a missionary from the local African Church, and we are given the opportunity to work in the townships, building playgrounds, repairing homes, taking the children out...etc. The children are always so happy and grateful, I feel very humbled by them. The townships are very interesting and the sense of community is incredible despite the terrible poverty. Fundraising is, of course, an important activity in the Children's Home and I have enjoyed liaising with the public for donations (especially at Christmas time) and helping at Golf and choir festival events.

I haven't had much time to travel since I got here but I did spend a week in Johannesburg visiting another Project Trust site. I have been to Robben Island (where Mandela was imprisoned), climbed Table Mountain the long way round - it took 6 hours! - and visited Cape Point (the southern most peninsula of Africa). I also plucked up all my courage and went shark cage diving and have been to some of South Africa's beautiful beaches. Before I go home in August I hope to spend 6 weeks travelling and camping in Namibia, Botswana and throughout South Africa. I have spoken to the guides organizing this tour about my diet already and I will be bringing my supplement and doing as best I can with the food available. I'm very excited!

Getting used to the Children's Home and South Africa in general hasn't been easy and there are many times when I want to go home, but there are just as many when I dread the day I will leave and wish to stay here forever. I've learnt so much from the children and from living and working in Africa. One thing that has definitely been highlighted is how lucky I am. PKU has been easy to deal with, thanks to the help and support of my family, friends, health professionals and SHS, and after seeing children with much more serious problems and afflictions I feel so grateful. There is no need for PKU to hold us back from anything.

I would like to say a huge Thank You to those of you who so generously supported my year. I can't mention names as I might leave someone out but you know who you are. I hope to catch up with you again at next year's PKU conference!

Clare Gibbings  
(19yrs and Classical PKU)



# Holidays

## – some personal experiences

Those of you who came to the conference this year will have heard Warren Knight talking about travelling with his 4 children, all of whom have PKU. He was inspirational in the variety of holidays he has tried and very helpful in how they as a family manage to do so much. I asked him for his top tips and here they are – practical and encouraging.

Sara Bartlett visited America with her daughter who has PKU and she did it a different way. Her account of using low protein products available in America is more expensive but that was a treat for her daughter as well as mother!

As Sara mentioned the use of American protein substitutes I have included information from SHS and Vitaflo on how to guarantee your normal protein substitute whilst on holiday or abroad for other reasons. This is the preferred option to having to use unfamiliar and perhaps not totally suitable product. You should always consult your doctor and dietitian before making any changes to your medication.

### Top Tips for Travelling with PKU Children

**TOP TIP:** Did you know you can request a special meal on the long haul flight and they will do their best to accommodate a suitable protein free meal. Alternatively you can ask for childrens meals and just let them pick at the bits they are allowed to eat.

**TOP TIP:** On long haul flights and certain short haul carriers, like British Airways – you can request extra baggage allowance as you have to transport medical supplies. Unfortunately, easyJet and Ryanair charge for hold baggage anyway and just charge you more for taking more.

**TOP TIP:** Use a hard case to hold your medical supplies and mark it clearly on the outside. This way you reduce any chance of the items being carried getting damaged.

**TOP TIP:** When travelling to the USA you should tick the box saying we are bringing food into the country on the white customs form. The customs officials usually look at the form and ask a couple of questions and then let you through. More often than not, the queue for the 'we have something to declare' is a lot shorter than the other queue.

**TOP TIP:** If you are visiting Disney World in Florida, in certain restaurants in each of the theme parks you can pre-order a PKU meal. I can't go into all the detail here but you need to contact Disney and fill out a *Walt Disney World® Guest Allergy/Dietary Needs Request Form*. You do need to know where you are going to be and when you are going to be there. But it doesn't half make things special.

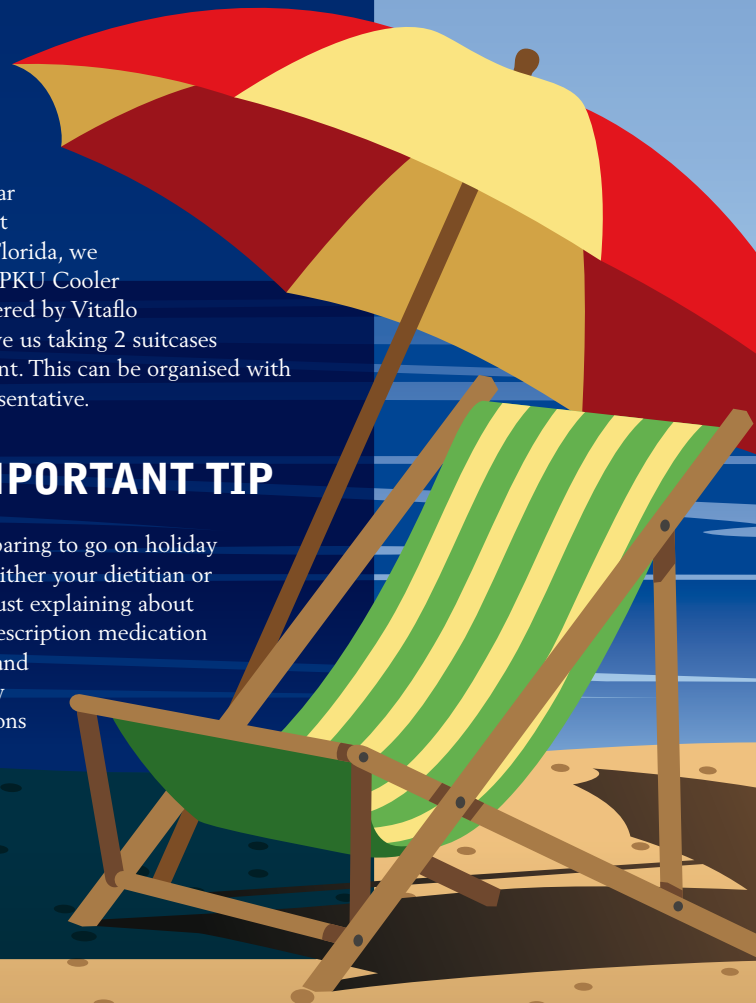
**TOP TIP:** There are now quite a few Apart-hotels, as they like to be called. So you get the benefits of a hotel but the comfort of an apartment. These Apartments come with a kitchen allowing you to prepare meals.

**TOP TIP:** We make sure we order enough PKU Cooler for the trip, well in advance of the holiday. But we also make sure there is at least two weeks left at home for when we return. We know how easy it is to be focused on the holiday and what you need for the trip and forget that it will take a couple of days at least to re-stock when you get home.

**TOP TIP:** This year on our trip to Walt Disney World in Florida, we are having all our PKU Cooler supplement delivered by Vitaflo USA. This will save us taking 2 suitcases of PKU supplement. This can be organised with your Vitaflo representative.

### MOST IMPORTANT TIP

**TOP TIP:** In preparing to go on holiday get a letter from either your dietitian or your consultant, just explaining about PKU and what prescription medication you need to take and always take a copy of your prescriptions with you.



# Are you planning a trip to the USA?

Our family has been fortunate enough to visit America over the past few years. Before our first holiday I scoured the internet in the hope of finding a source of low protein foods to buy so that whilst there our daughter could try something new and different. The company I discovered was Cambrooke Foods, contactable at [www.cambrookefoods.com](http://www.cambrookefoods.com) and we have tried many of their delicious products.

Helpful hints when making an order:

- Sign up and create a password.
- Look through the wide choice of foodstuffs and select. The scary part is that you need to order a minimum amount of cold-shipped or dry-shipped items. The 2 will be couriered separately. Dry ship is currently \$30 minimum and cold shipped, \$50.
- The postal charges will seem expensive if you order cold shipped items as they are carefully packed in dry ice and ice packs and transported speedily for obvious reasons. Be reassured that the on-line ordering process is straightforward and you will see the costs and charges clearly before you agree to buy.
- Send Cambrooke a note with your order telling them the date you are arriving at your accommodation – you don't want perishable goods hanging around for days before you get there. Give them enough notice and their delivery will be very efficient.
- Have a word with your accommodation to check that they are happy to accept the delivery and warning them when to expect it.

As we are lucky having so many foodstuffs available in the UK on prescription, at first sight ordering foods that you normally don't have to pay for seems expensive. For us the convenience of having low pro food on holiday that a) you don't have to cart in your suitcase and b) you don't have to prepare when you get there has been well worth the expense. I speak with the experience of cooking everything from bread in a wonderfully equipped mountain chalet to proving and cooking bread on the single hot plate in the corner of a Greek studio.

Also the opportunity to try different foods has been fantastic. We have particularly enjoyed Cambrooke's homemade breads, filled pasta, (meltable) cheese slices, chocolate raisins and cranberries, and heavenly chocolate brownies and peanut butter chocolate chip cookies (truly, they are delicious)!

NB. The camino-pro products are protein replacements which you will not be able to order without agreement and prior arrangement with your dietitian/consultant/GP. This isn't really an option when you are visiting for a short period and you would almost certainly have to pay for them. We can however vouch for the good choice and flavours available and wonder if the ready mixed products available in the UK would be stable if frozen into ice-pops, one of Cambrooke's tasty products?

One final and very important note is that all of the products come with information that includes their protein content. Some have enough protein that they need to be counted as part of your exchanges. Obviously you need to discuss this with your dietitian before trying. If you try Cambrooke Foods I hope you love them as much as we do,

Sara Bartlett

*All PKFoods are now available on prescription*

Chocolate Chip Cookies, Cinnamon Cookies, Orange Cookies, Pasta Specials, White Sliced Bread, Crispbread, Egg Replacer, Flour Mix, Cherry Jelly Mix, Orange Jelly Mix, amibex Cookies, amibex Biscuits, amibex Ruskis

UNIT 270 CENTENNIAL PARK CENTENNIAL AVENUE  
ELSTREE BOREHAMWOOD HERTS WD6 3SS  
TEL: 020 8953 4444 FAX: 020 8953 8285  
Website: [www.pkfoods.co.uk](http://www.pkfoods.co.uk) Email: [info@glutenfree-foods.co.uk](mailto:info@glutenfree-foods.co.uk)



# A Note From SHS and Travel Help in the USA

We would always recommend that the patient obtain enough product prior to travelling and travel with all the product required for the duration of their stay abroad. Should they run out for some reason the US have on occasions supplied to non-US residents. US products often have a slightly different micro-nutrient profile than products available in the UK and the rest of the world as they are formulated to US guidelines; therefore the products they receive may not be exactly the same as they do in the UK despite having the same or similar names.

There is a form which lists the products that the US supplies to their patients and this must be filled in by the patients dietitian. The patient will have to pay for the product as well as delivery. Prices are available from the SHS on request. They US office were unable to give specific prices when I requested but I have asked for an example of cost as a guide so I will forward this on when I receive.

The loprofin food range can be purchased for short term stays directly from the US office without a consent form in this instance all they would need to do is call 1-800-365-7354, Monday-Friday from 8:30am-5:00pm, EST and order the product they require. As long as they have the address of the hotel, there should be no issues. They can pay by credit card. Standard shipment would take about 1 week but they are able to fast track that shipment at an extra charge, this will depend on their location in the US.

It is essential to note that this should be used as a last resort. Patients should try to take all products needed with them to ensure there is no disruption in their diets.

Leigh can be contacted as below for further information and advice:

Leigh Garster, Medical Development Department, SHS International, 100 Wavertree Boulevard, Liverpool, L7 9PT

## Help from VITAFLO when travelling abroad

### Vitaflo Ltd Protein

#### Obtaining protein substitute abroad

Vitaflo Ltd can organise for protein substitute to be delivered to the address of your holiday destination. It is easier for them to do this if you are receiving your protein substitute via the home delivery system of Homecare, but it can be done even if you are not. Delivery is possible to most of modern Europe including Turkey and Greece and also to America, Australia, New Zealand and the

Scandinavian countries. Eastern block countries as well as China are more difficult. In the latter it can sit in customs for a year.

You or your dietitian should telephone Vitaflo's head office or email them giving as much information as soon as possible i.e. address of where staying, protein substitute and quantity. Vitaflo will receive confirmation that the protein substitute has been delivered by fax and will then confirm with the family or individual that it is there.

Vitaflo Head Office Number 0151-709-9020  
and [debbiejones@vitaflo.co.uk](mailto:debbiejones@vitaflo.co.uk)

## NSPKU MERCHANDISE



### NSPKU will be selling two designs of Christmas cards this year

Each packet contains 10 cards with envelopes and will retail for £3.00 per packet.

This message inside reads:

*With Best Wishes for Christmas  
and the New Year*

For more information please contact Mike Bailey, see inside front cover for contact details.

# Vitaflo and Homecare ...working together to deliver a better service

## STRESS FREE!

No need to order and pick up your prescription for your Vitaflo products from your GP – Homecare can do it for you!

## WORRY FREE!

Designated delivery date – all your Vitaflo products delivered to an address of your choice – home, work or even a holiday destination\*!

## HASSLE FREE!

Freephone helpline with extended opening hours for all your delivery queries – fitting into your busy lives at times that suit you!

## FRIENDLY!

Designated homecare coordinator who will liaise with you each month about your delivery!



Homecare is a FREE reliable and trust worthy home delivery service for all your Vitaflo products.

For more information or to register with this service please contact your dietitian.

**Homecare Freephone Helpline: 0808 100 1990**



&



**Homecare**

*...because life is complicated enough!*

\*Holiday destinations within the UK. Homecare is not available in Northern Ireland or the Republic of Ireland

# Report on the annual NSPKU Conference – Torquay 2009



There was quite a debate within the NSPKU about holding the annual conference so far south in Torquay. Our concern was that attendance might not be so good because of the distance for those in the Midlands and north to travel. What actually happened was really surprising and encouraging. There were 40 for whom this was their first conference and the distance did not deter those from Glasgow or Holland. There are always those who attend regularly but it was good to hear the first timers saying they would be making the effort to go again next year.

## So what draws them all in?

Well, for the children it is 'PKU-land' everything is for them (and the adults too with PKU) for they are the centre of the weekend. No-one has to worry about the diet (except me) and the hotel being completely ours for those 3 days means you can strike up a conversation knowing it is one big family.

We had some excellent speakers with a nice mix of professionals and the real experts- the patients and parents. As with most conditions we look for improvement or a cure. Two speakers explained new treatments; Dr Elaine Murphy and Dr Christineh

Sarkissian. Dr Murphy from the Charles Dent Metabolic Unit in London spoke about BH4 (tetrahydrobiopterin) which is synthetically produced and known as Kuvan. Kuvan helps the enzyme phenylalanine hydroxylase to work better

and it is estimated that 20-30% of patients who generally have some enzyme function will show some response. Kuvan is taken as tablets dissolved in water or juice and was approved for use in September 2008. However, it is a very expensive treatment (£50k for the lower dose and £100k per year for the higher depending on the patient's response.) Metabolic units plan to draw up guidelines for determining responsiveness and how it should be used in the treatment of patients.

Dr Christineh Sarkissian then spoke about the research and administration of injections of an enzyme substitution called PAL. This enzyme is made from a plant source and it is able to convert phenylalanine to trans-cinnamic acid and ammonia. If it is proved safe and effective during clinical trials (currently being undertaken in the US) then it can potentially be used to reduce phenylalanine.

The new dental leaflets written by Dr Sally Craig were launched at the conference with the statement 'Healthy Teeth make for a Happy Smile'. Dr Craig showed some graphic pictures of the problems caused by poor oral hygiene and diet in the general population and then explained how best to protect the teeth of those on the PKU diet. Traumatic injury, orthodontic treatment, topical fluorides, toothpastes, brushes and all practical aspects were covered in her helpful presentation. We all remember a lovely smile and this talk made us realise how important and precious it is.

Recently there has been an explosion of foods in the supermarket enriched with omega 3 fatty acids. The explanation of the essential fatty acids –omega 3 and omega 6 was amusingly and expertly portrayed as a Louis Walch versus Simon Cowell battleground. Simon Cowell being the dominant omega 6 and little Louis Walch the omega 3. In the PKU diet there is plenty of omega 6 fatty acids





but not much omega 3 and this imbalance limits the conversion of the omega 3 family to the longer chain fatty acid DHA. This DHA is important for brain development and function, for a healthy nervous system, fine motor skills, eye structure and clarity and many other important roles. The only direct dietary source of DHA is from fatty fish but of course the PKU diet does not include this food so, Dr MacDonald is taking part in a European study of children looking at the best optimal amount required in the PKU diet. For some time infant protein substitutes have contained DHA and newer substitutes e.g. Anamix Junior Liquid (SHS International) and the Cooler range (VitaFlo International) also have it added. It is possible to give DHA supplements but the type and dose should be checked with the dietitian. It should also be possible to make some DHA from the essential omega 3 fatty acid alpha linolenic acid. Rape seed, walnut, flaxseed and soya oils all contain some of this fatty acid. I think we shall be hearing more about this next year and Anita will be back to keep us informed.

I started working for the PKU Society over 21 years ago and I remembered the next speaker as a young boy of about 7 years old when his mother was treasurer of the Society. He came back as a married man, working in the police force, to talk about growing up with PKU and life on diet in work. Michael was one of the funniest natural speakers we have heard at a conference. He was inspirational; full of stories which made us laugh but at the same time highlighted the difficulties some children can face if they are different. For a start he has ginger hair and was described as the 'ginger freak who can't eat' by some at school but he said he had his close friends and his older brother and his advice was 'don't let them get you down. If they don't like it they are not worth knowing'. He has been a police officer for 2 years and loves his job. Keeping on top of rules and regulations is part of it and he does it, keeps to his diet and says his dietitian loves him! He reminded us of how much better the diet is today compared with when he was a child when even the ducks wouldn't eat his bread. He and his wife Philippa adapt recipes, invite friends round and live life to the full. His wedding day meal was based on what he could eat and I thought this was such an important point. He is living with someone who is totally supportive. He'll perhaps not thank me but I have to say his parents would have been so proud to hear him.

Another speaker who knows very well what family life is like was Warner Knight. He described his family thus 'I have 4 beautiful children all of whom have PKU'. Warner has his own travel company and whilst he spoke a continuous slide show of places they have visited rolled on behind him. He said the key to successful holidays is plenty of research and preparation. 'Last minute breaks we can't do'. I have asked Warner for his Top 10 tips as he had so many good ones. Look out for these in News and Views the Society magazine and on our web site.

Keeping up to date with what's useful and new in the supermarket is becoming an annual feature. This year our table was set with some unusual and some practical foods. Delegates were asked to bring along something to be displayed and then we talked about them. The diet choice gets wider and more interesting and this feature table of items helps us all pass on the ideas.

The trade stands were welcoming and I think the companies enjoy meeting their clients to have some feedback. There were competitions to win some fabulous prizes, perhaps the child going home with the Wii more excited than the child who won the bread maker, but she will love it long after the Wii has been overtaken by the next game!

Children's outings, learning about PKU in the crèche, decorating cakes and lollipops, quizzes, raffles and having a drink in the bar all made for a really friendly weekend. The food was super, the sun shone brightly and questions were asked by many of those first timers where it would be next year. I'll keep you posted.

#### Pre-Christmas Long Weekend in Torquay Friday December 18th to Monday December 21st.

The Rainbow Hotel is putting on a long weekend for families and adults with PKU. The diet will be available and all the hotel's facilities. The cost is really good value so you might like to think about a break with the family just before Christmas and enjoy the diet without having to do it yourself.



## Thanks to our supporters

We have once again been very fortunate with our sponsors and I would like to take this opportunity to give our thanks to:

**SHS** - who provided £16,000 to cover the costs of assisted places, the children's and teen's trips at last year's conference and for our ESPKU costs.

**VitaFlo** - who have provided a total of £6,000 towards the costs of Publications.

We would also like to thank; Eileen and Martin Green of Fate Special Foods, Steve and Linda Fletcher from FirstPlay Dietary Foods, Ellis Ward at PK Foods, Don Lewis of Ultrapharm and David Green of General Dietary for their foods. These have been donated generously not only for the conference but at all the events held during the year. Their support helps us hugely in our work and makes the events' meals tasty and special!

I would like to pass on our thanks to everyone who has supported the NSPKU throughout the year; especially to Lindley Methodist Tuesday group who raised £1,350 and RAF Uxbridge for their donation of £1,250, not forgetting those who continue to support us via regular direct debit or workplace schemes.

# Can someone please explain to me what PEG-PAL is and how it works?

This question came to us via the PKU community and prompted the following response:

Christineh N. Sarkissian, John Adams and Alejandra Gámez<sup>1</sup>

## WHAT IS PAL:

Phenylalanine ammonia lyase (PAL), is a protein, an enzyme, found in plants, fungi and bacteria, that converts L-phenylalanine to *trans*-cinnamic acid and ammonia (1, 2).

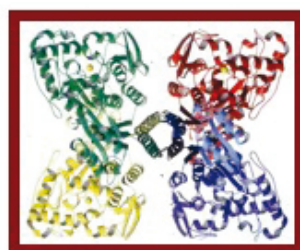
## WHY WAS PAL THE CHOICE OF ENZYME AND HOW DOES IT WORK:

PKU and related hyperphenylalaninemias result from compromised phenylalanine hydroxylase (PAH) activity, causing excess phenylalanine (Phe) accumulation (3). The harmfully augmented levels of Phe can be diminished by phenylalanine metabolizing enzymes; via *enzyme replacement* with PAH or *enzyme substitution* with PAL.

Enzyme replacement therapy with the native PAH (the primary choice), requiring a multi-enzyme complex to function, presents a number of challenges. PAH is inherently unstable; its complex activity and cofactor (BH<sub>4</sub>), oxygen and iron requirements, in addition to the inherent degradation sensitivity and potential immunogenicity in a person lacking the functional enzyme, further complicates its therapeutic viability (3). PAL is a more stable enzyme and does not require a cofactor to function. Therefore enzyme substitution therapy with PAL alone could metabolize Phe. PAL enzyme can act as a surrogate to the deficient PAH enzyme and convert the excess phenylalanine to *trans*-cinnamic acid and ammonia. *Trans*-cinnamic acid is a harmless metabolite, as it degrades and is rapidly excreted in the urine as hippuric acid, and the ammonia does not accumulate in sufficient quantity to pose a threat of hyperammonemia (1, 2, 4-6).

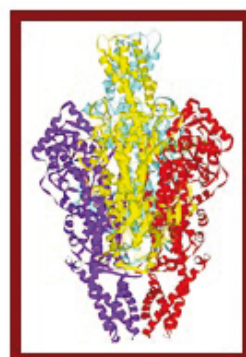
<sup>1</sup> Sarkissian and Gámez are research scientists from the Departments of Biology, Human Genetics and Pediatrics, McGill University-Montreal Children's Hospital Research Institute, Canada, and Centro de Biología Molecular "Severo Ochoa", Universidad Autónoma de Madrid, Spain, respectively. With over two decades of combined research experience in the field, the main focus of our research has been the development of an enzyme substitution therapy with PAL for the treatment of PKU. John Adams is a PKU parent and the President & CEO of the Canadian PKU and Allied Disorders Inc.

### 3-D crystal structures:



Phenylalanine Hydroxylase  
(PAH)

*Courtesy of Dr. R.C. Stevens' Laboratory.*



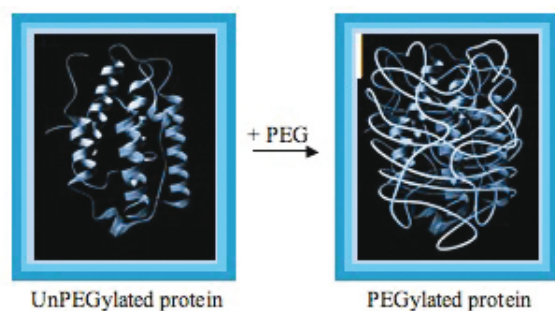
Phenylalanine Ammonia Lyase  
(PAL)

## WHAT IS PEG:

PEG stands for polyethylene glycol. It is a water soluble and non-toxic compound that is safely cleared from the body. Its primary use to date in currently approved drugs has been to favourably alter the properties of biologics (7). The attachment of PEG to a therapeutic protein (a process called PEGylation) can improve its biopharmaceutical properties by increasing its stability and resistance towards inactivation and directed degradation. It also diminishes immunogenicity (ability of a foreign protein (drug) to elicit an

immune response), by masking the presence of the drug from the host's immune system, and enhances and extends its performance by reducing clearance and prolonging circulatory time (8).

#### Schematic Drawing of PEGylation Technology:



[www.nektar.com](http://www.nektar.com)

## WHY IS PAL PEGYLATED FOR THERAPEUTIC USE:

An injectable form of phenylalanine ammonia lyase could have a therapeutic profile for PKU. However PAL is a non-human protein that the immune system recognizes as foreign. Long-term reduction of phenylalanine levels by PAL is hampered by clearance of the enzyme via an immune response; PAL alone (without PEGylation), although shown to be harmless upon repeated dosing to the PKU mouse model, permanently loses its ability to metabolize systemic phenylalanine one week following initial administration (2).

PEGylation of PAL controls its immunogenicity. Modification of primary amino groups (largely confined to the protein surface of PAL) by attachment of activated forms of PEG (producing PEG-PAL), reduces and nearly eliminates the recognition of the now tailored enzyme by the immune system, and therefore prolongs its life while retaining its function (6).

## RESEARCH:

We have shown that the blood phenylalanine concentration in the PKU mouse model can safely be reduced down to normal levels by the administration of PEG-PAL. All PEG-PAL formulations that we tested were effective catalysts; they had prolonged activity and certain forms had profoundly suppressed immunogenicity, a desirable feature for clinical use (2, 4).

Applied to humans with PKU, this type of substitute enzyme therapy could provide substantial relief from the rigours of the diet and improve compliance, metabolic control, and clinical outcome in this relatively frequent disorder (2).

Phase I clinical study of PEG-PAL for the treatment of PKU was initiated on May 20th, 2008 (9).

## ACKNOWLEDGEMENTS:

These developments would not have occurred without the contributions of Drs. Charles R. Scriver, Raymond C. Stevens, Lin Wang and all our colleagues at BioMarin Pharmaceutical Inc.

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# Fate Special Foods recipes



*Hello,*

*These bread sticks are useful to have as a crunchy alternative to any other type of bread and really good served with a suitable dip. They are ideal to pack into a picnic or lunch box. They will keep for a few days in an airtight container, but they are best stored in the freezer, as they only take a couple of minutes to defrost.*

*The method uses a piping bag and nozzle to shape the breadsticks. It is best to use a large nylon one available from cake decorating shops, and a plain nozzle just under a centimetre in diameter*

## FATE BREAD STICKS

### Ingredients

250g Fate All-Purpose Mix

½ tsp salt

½ sachet yeast (enclosed with the mix)

15 mls oil

200 mls warm water

Pre-heat oven Gas 4. 175°C (350°F)

### Method

Place the Fate All-Purpose Mix into a bowl, and stir in the salt and the yeast. Put the measured oil and water into a separate jug. Have ready an electric hand mixer and pour the oil and water all at once onto the dry ingredients. Immediately start mixing to get everything blended together. Then mix on slow speed for about 30 seconds until smooth and thick.

Place a nozzle into the base of the piping bag, and place the bread stick mixture inside the bag. Twist the top of the bag tightly against the mixture.

Lightly grease a baking tray, and pipe lines of mixture leaving a small space in between each one. When the tray is full, dip a knife into water, and neaten the ends if necessary. Place the tray into a large polythene bag, and put into a warm place to prove. Leave until risen about half the size again.

Place into a pre-heated oven, till risen, brown, and crisp. This may take anything from 20 mins to 40 mins. Remove to a wire rack to cool.

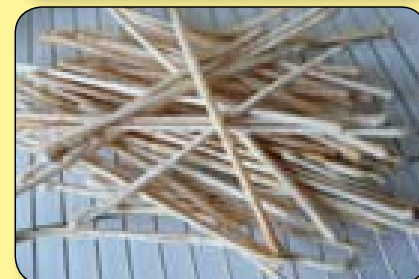
### Note:

The cooking time may vary greatly for different ovens. The first time you make them, check them every five minutes.

They need to cook steadily, so that they turn golden brown, and also become crisp all the way through.

It is best that they do not brown too quickly.

As a change, you can make bread sticks in different sizes and lengths. Make shorter ones ~ ideal for younger children or use a narrow nozzle and make long elegant thin ones. Just remember to vary your cooking time depending on which size you do.





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



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

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Fate Low Protein Cake Mix

Fate Low Protein Chocolate Cake Mix



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# Vegetable Bolognaise



## Ingredients:

15mlsp (1tbsp) olive oil  
1x red onion, chopped  
1x large garlic clove, crushed  
100g carrots (about 3), finely sliced  
1x red pepper, deseeded and chopped  
75g mushrooms, chopped  
500g carton creamed tomatoes  
50g broccoli, chopped  
Handful fresh basil leaves  
250g Loprofin Spaghetti

## Method:

1. Heat oil in large pan, add onion and garlic and fry for few minutes to soften but not colour. Add carrots and cook 5 mins, then add pepper and cook for further 2 mins. Stir in mushrooms and creamed tomatoes, bring to boil. Add broccoli and basil and simmer 5 mins or until all vegetables are tender.



2. Whizz vegetables in food processor, in batches, until very finely chopped. Return sauce to saucepan to warm through.

3. Meanwhile, bring large saucepan of water to boil and cook spaghetti according to packet instructions.

Drain and toss through sauce to serve.



# Vegetable Moussaka



## Ingredients:

1x batch Vegetable Bolognaise Sauce  
4x 15mlsp (4tbsp) olive oil  
1x jar white lasagne sauce  
Small aubergine, sliced  
2x 15mlsp (2tbsp) Loprofin breadcrumbs  
1 pkt crushed Quavers (optional) 1/2 exchange per small pkt  
Oven temp: 190°C /375°F/Gas Mark 5

## Method:

1. Make a batch of bolognaise sauce.
2. Heat oil in frying pan and lightly fry aubergines on both sides for 1-2 mins, put to one side.
3. Spoon some sauce into ovenproof dish and cover with layer of aubergines and then layer of white sauce.
4. Repeat, ending with white sauce.
5. Sprinkle top with Loprofin breadcrumbs and Quavers.
6. Bake in oven for 25 mins until golden brown

# Cottage Pie



## Ingredients:

1x batch of Vegetable Bolognaise Sauce  
500g parsnips/sweet potato/swede/turnip (or a mixture of these) peeled and chopped  
25g butter  
Salt and pepper  
25g Loprofin Breadcrumbs  
1 pkt crushed Quavers (optional) 1/2 exchange per small pkt  
Oven temp: 190°C /375°F/Gas Mark 5

## Method:

1. Boil prepared vegetables in salted boiling water until soft.
2. Drain and mash with butter, salt and pepper until smooth.
3. Spoon bolognaise mixture into ovenproof dish and cover with mashed vegetables.
4. Sprinkle top with Loprofin breadcrumbs and Quavers.
5. Bake for 25 mins until golden brown.



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# Festive break at The Rainbow Hotel

Following this year's successful and enjoyable weekend conference at the Rainbow Hotel in Torquay, Devon, we've teamed up with the hotel to offer an exclusive break for readers and their friends and family. If you'd like a short break away, to enjoy the friendship and support of other PKU families, read on!!

The Rainbow Hotel has 140 bedrooms, several of which are family rooms for 2 adults and up to 3 children. All the rooms are en suite, with tea and coffee making facilities and flat screen TVs. There is a Health & Leisure Suite, incorporating heated swimming pool, steam room and mini gym, all free of charge and open from 7 am to 7 pm.

The weekend 18th – 21st December has been offered at an exceptional rate exclusively for PKU families. The offer was made at this year's March conference, and was received enthusiastically by many families, but the hotel need 80 adults to ensure the weekend can go ahead. Although details of this break will only be circulated via us, the Rainbow are happy for friends and family to attend, the whole hotel is available, and the more the merrier!

The weekend starts on Friday, with a drinks reception early evening in Club Rainbow, followed by 3 course Carvery Dinner. Full PKU menus are as important as the carvery on this weekend – and executive chef Roger will recreate the dishes that we enjoyed on the March conference. (See menus). . The hotel will be fully decorated for Christmas, and the Salvation Army Band will enhance

the atmosphere with well-loved carols. A disco with music for every taste will be the entertainment in the Club on your first night.

Full English Breakfast (again with PKU options) and 3 course dinner on Saturday, with an evening of entertainment in the club, or the opportunity to relax in the Empress Ballroom with a drink and catch up with old friends.

On Sunday evening enjoy a great Christmas Show, with all your favourite Christmas songs, and some dancing and comedy.

If you're driving to the hotel, it's easily located from the M5 motorway – or Torquay train station is nearby and National Express service are options. There is free car parking on site and there are several very worthwhile places to visit. The hotel can offer discounts at local attractions such as Kents Caverns and Babbacombe Model Village.

The cost of the weekend is just £75 per adult, and children between 3 and 14 stay free if sharing their parent's room, paying only a £10 meal plan for the whole weekend. Children under 2 are free of charge.

For more details of the hotel, visit [www.rainbow-hotel.com](http://www.rainbow-hotel.com) If you're interested, please call the Rainbow on **01803 213232**. No deposit is required, but the hotel would like to know whether there will be enough support for the weekend to go ahead.

## NSPKU FESTIVE BREAK

**Friday 18th - Monday 21st Dec 2009** 3 nights

- Fully Decorated Hotel
- Welcome Drinks Reception
- Traditional Turkey Dinner with all trimmings
- P K U Menus all Weekend
- Nightly Entertainment & Music for Dancing
- Carols with Salvation Army Band
- Special Christmas Cabaret Show
- No Single Occupancy Supplement
- "All you can eat" Breakfast & Carvery Dinner

A special Festive Break exclusively for NSPKU Members, their friends, family & groups  
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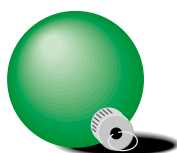
**Just £75.00 pp for 3 nights!!!**

Children sharing parents room stay FREE, £10 weekend mealplan applies

*Go on, treat yourself and your loved ones to a  
well deserved pre-Christmas treat!*

Quote PKU Festive Break & book now on 01803 213232

**[www.rainbow-hotel.co.uk](http://www.rainbow-hotel.co.uk)**



# Dietitian's Report



## Making a Chocolate Cake a bit special

I recently received a query from a dietitian about Betty Crocker cake frostings and buttercream. A patient had mistakenly thought that one of the chocolate flavours was a 'free' food and had been enjoying rather a lot of it. The question was 'What is the protein content of them all and was it the reason blood levels had gone up'?

There is no nutritional information on the Betty Crocker range but the customer relations department of the company is very helpful and quickly provided me with the help needed.

So here is the information:

**Buttercream Style Vanilla** 0.02g protein per 100g  
- FREE

**Rich and Creamy Vanilla** 0g protein per 100g  
- FREE

**Rich and Creamy Chocolate Fudge** 2.20g protein per 100g. **So this one must be counted.**  
45g = 1 exchange

**Buttercream Style Chocolate** 0.46g per 100g.  
So 200g = 1 exchange

This is a lot of buttercream and is plenty to fill a 18cms (7 inch) chocolate cake and to have enough to put on the top too. This is what I have done in

the picture. There are about 8 slices to this cake so you could have one or two slices and not count it.

I melted some low protein 'chocolate' bar (use either Duobar or Vitabite bar) for 1 minute in an uncovered bowl in the microwave and then filled some little heart moulds and placed in the fridge to set for 20 minutes. You can get all kinds of shapes of moulds and a good web site is [www.homechocolatefactory.com](http://www.homechocolatefactory.com) or [www.cakecraftshop.co.uk](http://www.cakecraftshop.co.uk). You could also just grate a little of the low protein chocolate bar over the cake which looks attractive.

Raspberry jam in the centre of the cake as well as the buttercream filling is also really nice. Chocolate flavour and raspberry go well together.

The FATE chocolate flavour cake mix and SHS Chocolate flavour cake mix are easy to use. Just make sure you use the right size of tin.



## What's new and really useful in the Supermarket?

At the conference we had a table of new foods which people have found and which they find really helpful in making the diet easier and more interesting. I am featuring a few of these things now but do look out yourself and let me know about anything you find. I can keep on featuring them to help everyone.

### Morrisons

#### Zingy Ginger Sauce

Free

- Brush over vegetable kebabs before grilling
- Add to stir fries and heat through before serving
- Pour over sorbet or Loprofin dessert

#### Full Flavoured Sundried Vegetable Mix

Free

- Add to bread mix to make loaves, pizza bases or rolls
- Add to pasta during cooking to change flavour of pasta
- Add to vegetable casseroles
- Add to pasta sauces to change the flavour
- Add to rice while cooking

#### Delicately Fragrant Tomato and Rosemary Mix

Free

All of the above plus

- Add to pastry



#### Air Dried Orange Peel Granules

Free

- Add to pastry for a sweet pastry along with a pinch or two of mixed spice
- Add to cakes, muffins or biscuits
- Use in stir fries with honey, balsamic vinegar or brown sugar
- Use in a marinade for grilled vegetables





### Air Dried Lemon Peel Granules

Free

All of the above plus

- Sprinkle over roast potatoes or vegetables along with olive oil, salt and ground black pepper, fresh herbs
- Add to olive oil and vinegar and use as a salad dressing
- Add to mayonnaise and use with salads, dips or baked potatoes



### Bank's

Cassava Vegetable Chips

These crisps are made from cassava which is very low in protein and they can be classed as free. If any crisp is made from cassava and/or tapioca and all the other ingredients are free then the crisps are free.

Thank you to Louise Brennand for the following food finds:

## Marks and Spencer

### Vegetable Oriental Selection

These can be found in the fresh and frozen sections of the shop. The frozen are quite a bit cheaper.

There are vegetable spring Rolls, Mushroom and coconut Wontons and Vegetable Cones.

They should be counted as follows:

Spring Rolls 1 roll = 1 exchange

Wonton 1 wonton = 1 exchange

Vegetable Cone 2 cones = 1 exchange



### Kellogg's Rice Krispie Squares with sugar coated pieces

These are another version of the plain squares.

1 square = 1 exchange



Louise also asked me about some flaked rice she found in Tesco's with a protein declaration of 0.1g per 100g. Too good to be true? Yes because ordinary rice is around 6 to 7g protein per 100g. If you do find something which you are a bit suspicious about take it to your dietitian to check before you use it. I have alerted Tesco's to this error.



## Rice Milk and PKU

Following the Food Standards Agency's warning about rice milk and its arsenic content the NSPKU advises the following:

Rice milk should not be given to children under 5 years old

For children age 5 years and over and adults it can be used in tea/coffee, on cereals and in puddings to a maximum of 300-400ml per day.

As an extra precaution pregnant women should also avoid rice milk.

Alternatives to rice milk are:

Duocal – liquid and super soluble powder

Calogen

Sno-Pro ½ exchange per 200ml carton

Loprofin drink ½ exchange per 200ml carton

Milupa lpd ½ exchange per 200ml

Oatly 'milk' 125ml = 1 exchange

Oatly Cream 200ml = ½ exchange (carton size 250ml)

60ml Double Cream = 1 exchange (can be diluted down to taste)

Rice grains and baby rice can be used as normal remembering their protein content so count as exchanges.

## Haribo Halal Gummy Sweets

It has been brought to my attention that some of the Halal sweets do contain gelatine. Please check carefully that the sweets you buy contain Gellan and not gelatine. Gellan is a synthetic form of gelatine and contains no phenylalanine.



## Advice about casserole cooking

*I received an email from the parent of a PKU child which went as follows:*

'This may sound a silly question but here goes. Many of the cook in sauces e.g. curry, chicken tonight etc are free foods for my child. If I make a meat curry or casserole can I spoon the sauce and vegetables from the same cooking pot on to my child's plate or does the cooking process release protein from the meat into the sauce and vegetable?' Both of us work so it would be a lot easier to do this and my child could eat more of less the same as us.'

**My reply:** You are correct about the meat releasing juices into the cooking sauce. As meat cooks it shrinks and does release high protein juices into the sauce. Traditionally these juices are used to make gravy and add a lot of flavour when roasting meat but the same will happen in the casserole. Also in casserole cooking the meat is cooked to be very tender and the test to see if it is cooked enough is to see if the meat flakes easily. This will undoubtedly happen in the casserole and little pieces of meat will be in the sauce. Meat is extremely high in phenylalanine so it is best to cook the vegetables in the sauce in a separate container. You could make up several helpings and freeze some for another time.

Have you tried FirstPlay sausage mixes and Burger mixes made into little balls, fried and then added to the casserole mix for you child? Burger Mix 1 sachet = 1 exchange (both flavours) Sausage Mix 1 sachet = ½ exchange (all flavours)

## Dietary Information Booklet 2009/10

In the last issue of News and Views you received your new booklet. It is yellow this year. Changes were made to the way we count 3 vegetables as a result of doing some amino acid analysis. These 3 vegetables all have similar amounts of phenylalanine and in order to be consistent we made the changes. Those vegetables are:

Brussels Sprouts  
Broccoli  
Cauliflower

They contain a significant amount of phenylalanine and cannot be given freely but the amount is not high enough to warrant them being exchange foods. They have all been placed in the section on page 5 of the yellow booklet 'Restrict to ONE serving of any ONE item per day'.

Unfortunately cauliflower was missed off the list on page 5. I realised my mistake but the booklet was already being printed. It is included in the booklet up on our web site but you need to write it into your paper copy. I do apologise for this and any confusion it has caused but I'm also very pleased so many of you read the booklet carefully. I shall ask a few of you to help me with this job next time – it would really help as when you read something many times you can become too familiar with it.

## Sorbet in a Pot

These little pots of sorbet can be found on the shelves (not frozen) and are useful to have in the house for a pudding or snack. They are made with water and fruit puree and are free.

## Pink Panther Wafers

My 2 year old son Innes has PKU, and when I was shopping in Tesco last week my partner spotted these biscuits. They are gluten free/wheat free pink panther wafers. They have 0.01g protein each and 0.1g protein per 100g. We found them in the aisle with Tesco's "Free From" range. The list of ingredients are ; Flavoured filling (vegetable oil, sugar, beetroot red colour, natural flavouring), wafer (potato starch, vegetable oil, maize starch, stabiliser) Beetroot red colour, raising agent (ammonium hydrogen carbonate) and emulsifier (soya lecithin). I know there are prescription



wafers, but I just thought it was good that we can buy biscuits for him. I am sending you a picture of the wafers.

Mhairi Campbell

# A doubleblind randomised crossover trial of previously untreated people with Phenylketonuria

*Professor Glynis Murphy, Dr Philip Lee, Dr Brian Fitzgerald, Lesley Robertson, Allayne Amos, Pat Portnoi, Rosemary Hoskin, Maggie Lilburn and Eleanor Weetch*



Front Row: Allayne Amos, Lesley Robertson, Prof. Glynis Murphy

Back Row: Eleanor Weetch, Rosemary Hoskin, Dr Phil Lee, Dr Brian Fitzgerald, Pat Portnoi, Maggie Lilburn

In this News and Views is a report from Lesley Robertson who was the research dietitian for the Untreated PKU Trial. Screening for phenylketonuria was introduced nationally in 1969 and those born before this time did not receive the benefit of treatment of a low phenylalanine diet. High blood phenylalanine levels left untreated from birth causes a high risk of very difficult symptoms including mental retardation, eczema, body odour, sleep disturbance and self harm as well as harm to others. This results in isolation and problems of management for families and carers. Quality of life for the person with untreated PKU and those around them can be poor.

There have been several anecdotal stories of previously untreated people with PKU being helped by a low phenylalanine diet and Rosemary Hoskin, a dietitian working with these patients, had had success with treatment. She brought dietitians together for a meeting in 1997 to develop more interest and to learn from one another's experiences. From that beginning and some 6 years later a clinical trial got underway to establish if the diet was of benefit. This trial was the first double-blind randomised control crossover trial ever carried out with this group of patients. The results have been eagerly awaited by the metabolic world.

The 4 year trial has been expensive and it would not have taken place without the very considerable support of the Wellcome Trust and SHS International Ltd. The NSPKU also supported the work as the Society is there to help treated and untreated people and their families.

Here then are the results of that research work.

Please contact me if you wish to discuss a patient's treatment or would like a copy of our booklet Dietary Treatment of the Untreated Adult PKU. (small cost of £2 to cover costs – cheques made payable to NSPKU)

## Adults with previously untreated phenylketonuria (PKU) – a randomised control trial.

### Introduction

People with phenylketonuria, (PKU) born before newborn screening was introduced, were mostly left untreated. The majority now live in community care homes for people with learning disabilities (LD). Anecdotal reports suggest phenylalanine restricted diet can improve quality of life in this group. Following the identification of approximately 100 such adults, 36 participated in double blind randomised control crossover trial to assess the efficacy of a low phenylalanine diet.

### The Study

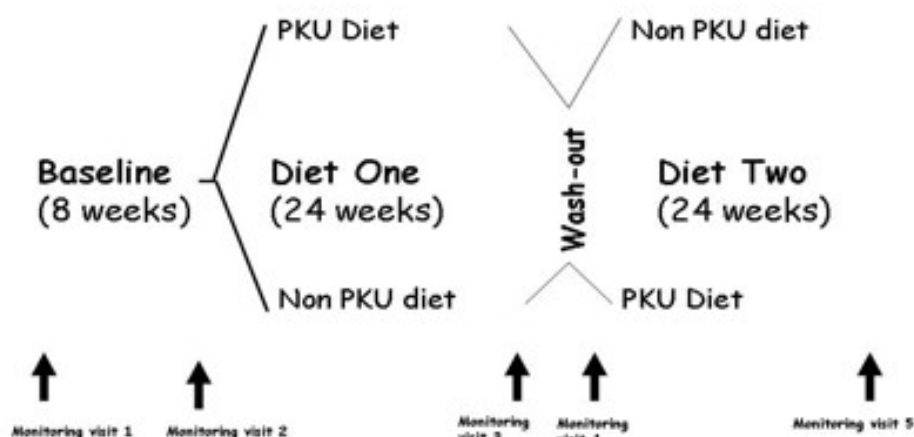
The 36 participants were each studied over a period of 60 weeks. Initially there was an 8 week baseline, after which the recruits were randomised into one of the two trial diets (low or normal phenylalanine diet) for 24 weeks. Between each period, there was a four week washout period in which normal diet was eaten. The final 24 weeks was the second trial diet.



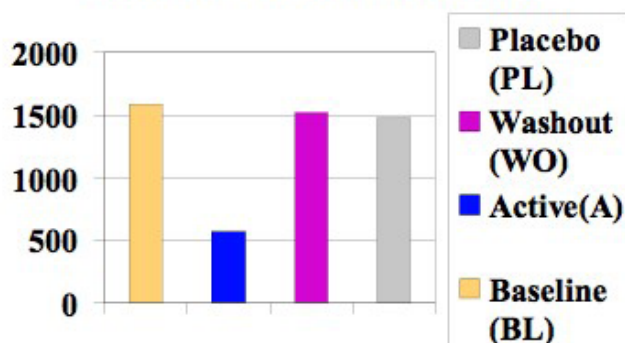
All were monitored at regular intervals throughout the 60 weeks, this included weekly blood phenylalanine and a range of behaviour monitoring including daily behaviour diaries, standardised questionnaires and video diaries.

## Results

Of the 36 participants, only 17 completed the full 60 weeks. 13 withdrew in the baseline or diet 1 due to the dislike of the amino acid product or blood test refusal. Three of the people who withdrew from diet 2 did so to swap to a PKU diet following a dramatic improvement on the first diet.



### Blood Phe for 17 completers



questionnaires and 3 short videos per visit. The analysis of these showed no significant differences during the active diet.

A daily behaviour diary was completed by care staff. This recorded 3 positive, 3 negative behaviours and carer comments. No significant differences were noted with positive and negative behaviours. However carer comments showed a significant benefit in the active phase. 240 comments were rated and are summarised below.

Comments in the active diet were significant when compared with the placebo and washout,  $P < 0.001$

## Conclusions

A low phenylalanine diet is difficult to institute in this group of individuals for a variety of reasons. However, the blinded, placebo-controlled nature of the study has been able to demonstrate meaningful benefits to this group of severely learning disabled adults, with 12 of the 17 subjects (71%) who completed the full 60 weeks continuing on a low phenylalanine diet.

## Acknowledgements

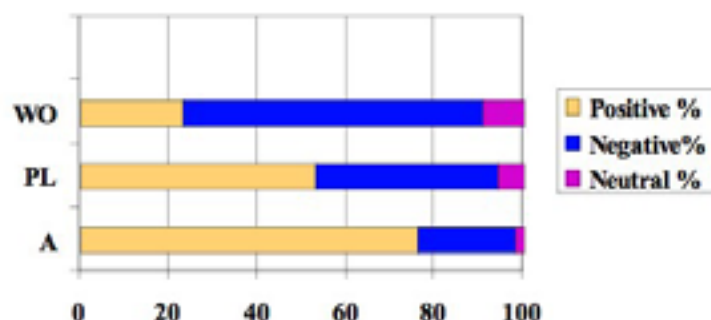
We are very grateful to the Wellcome Trust, SHS International and the NSPKU for funding, and the carers for their participation.

## Blood Results

The following table shows the blood results for the 60 weeks and demonstrates good phenylalanine control during the active low phenylalanine diet.

## Behaviour Results

Behaviour monitoring was carried out 5 times during the study – at the beginning and at the end of each phase. This consisted of standardised



# NSPKU 37th Annual Family Weekend & Conference 2010

## The Rendezvous Hotel Skipton

*Friday 5th March to Sunday 7th March 2010*

### Venue:

This year's NSPKU annual family conference and holiday weekend returns to Rendezvous Hotel in Skipton North Yorkshire. This hotel has proved popular with regular attendees and provides us with a delightful location to hold our event.

Escape from the pressures of city life to the picturesque North Yorkshire Dales and Herriot country and enjoy the facilities of one of the most versatile hotels in the area. Stunning views of the Yorkshire Dales from the peaceful waterside setting and lots of real Yorkshire warmth. The Dales offer some of the finest scenery in England; wild, tranquil, timeless landscapes dotted with grand castles, bustling market towns and beautiful gardens. If you simply want to find a quiet country walk it is all here!.

The Rendezvous Hotel, Skipton is very accessible from all parts of the country.

### Accommodation:

There is a great choice of bedrooms, from executive suites and family rooms to luxurious rooms for two, all offering you what you would expect from a top hotel. Comfortable, spacious bedrooms all with private bathroom, direct dial telephone, TV, hairdryer, trouser press, hospitality tray and individual controlled heating. Add all the hotel's business facilities and function rooms, and you will wonder why you had not enjoyed all the benefits of the Rendezvous Hotel before.

### 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:

80 en-suite bedrooms, training centre, 7 meeting rooms, The Papa Swan Bar, The Baby Swan Restaurant, 120 car parking spaces, indoor swimming pool, children's pool, whirlpool spa, steam room & sauna, solarium, gymnasium. (please note that due to the limited number of bedrooms available, a number of overspill hotels very close by to the Rendezvous Hotel, Skipton will be used to accommodate our guests. In order to maximise your chances of staying in the Rendezvous Hotel, Skipton, early booking is recommended. Guests who stay in other local hotels will conference and take luncheon & dinner in the Rendezvous Hotel, Skipton).

### 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.



The National Society for Phenylketonuria (United Kingdom) Limited



### Location:

**Air:** Leeds/Bradford airport – 30 minutes.  
Manchester airport – 1 hour 15 minutes.

**Rail:** Skipton station – 1 1/2 miles.

**Road:** The hotel is situated on the A629 Skipton to Keighley road 1 mile south of Skipton town centre.

### 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 (18 years and over)	£180.00
PKU Adult (18 years and over)	£90.00
Non PKU Child (11 - 17)	£90.00
Non PKU Child (4 - 10)	£45.00
PKU Child (4 - 17)	Free
All under 4's	Free

### Day Visitor Fees:

Non PKU Adults (17 years & over)	£35.00
PKU Adults (17 years & over)	£17.50
Children (0-16 years)	£10.00

### 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 of £30.00 per adult (£15.00 per child under the age of 12 years) with a £15.00 single occupancy supplement. This rate is for Thursday or Sunday night.