NSPKU News & Views
The National Society for Phenylketonuria (UK) Ltd
Issue 110

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In Touch

The Council of Management

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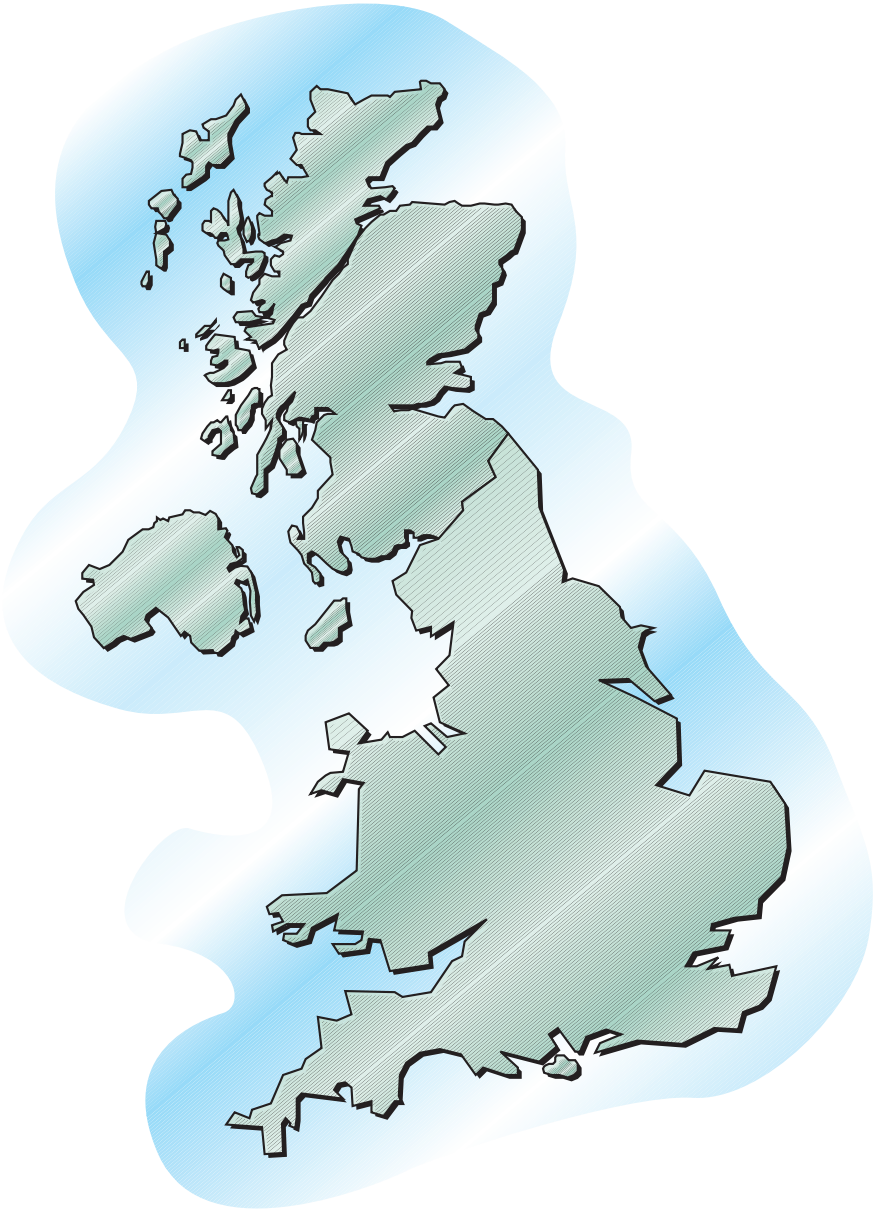

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Editor's Report...

Dear All

It is with a heavy heart that I edit my last issue of News & Views. I have been Editor for 13 issues of the publication and enjoyed each and every one. Kiri Thomas will now be taking over the rudder and I know she has further plans to keep the magazine up to the high level of quality it now enjoys. I wish her luck in her new role.

A fond farewell to all of the suppliers and manufacturers with whom I have enjoyed a great relationship and who have supported the magazine with badly needed advertisement revenue to help us pay our way.

And more so, good-bye to you lot, the readership, sponsors, and athletes/daredevils who get up to all sort of nonsense to support the coffers of the Society. Keep the letters rolling in. In such a small community of common sharing of difficulty (the diet and its administration) it's nice to know that you aren't alone and that until there is a permanent cure for PKU there will always be support in the form of the NSPKU.

Check out the article from PKU News – 'Recent Advances in Cell and Gene Therapy for PKU' the American Magazine. This is a fantastic article on possible future hope for the treatment of PKU. Many thanks to Virginia Schuett (Editor), who compiled the report and gave her permission for the reproduction of the article. Virginia has also been a great source of information, inspiration and help in previous issues of News & Views.

Thanks also to Keystroke Mill and Action Press for their input and to John McKenzie for having to print more labels for the mailing than was actually ever needed!

This is me. Pete B. Signing off.

Pete Bramley
Editor

Treasurer's Report...

I find I must begin this report with an apology. During the year we have been working hard to ensure that all our members have registered with the Charities Aid Foundation as we will be closing our membership list and using theirs very soon. We have therefore sent a number of letters to those who haven't appeared on their list yet. There was a slight hitch during the summer when I mischecked some of our names and as a result some of you who are fully paid up and are on CAF's list had a reminder. I take full responsibility for this mistake and am sorry for the confusion it has caused. If you are in any doubt as to whether you are on the list, please contact either CAF on 01732 520 00 or myself.

I should also point out that my contact details have changed so make sure you look me up in this issue rather than an earlier one. For any of you that have recently sent something to my old address, I do have a redirection in operation so this shouldn't be a problem for a while yet.

Another point I should mention on the subject of my old friends CAF is the requirement for gift aiding donations. We are only able to reclaim tax that has been **paid by the donor** on any donation. Therefore money raised by fundraising events and sponsorship will not be covered by a gift aid declaration form for the whole sum. Individual donors may sign a form for **their particular element** on which they have personally paid income tax. The gift aid declaration forms will hopefully be available on our website by the time you read this, or you can get one from me or our administrator, Lucy. They can be photocopied if you need extras. We both also hold copies of sponsorship forms and direct debit mandates for anyone who wants to donate on a regular basis, even if it's once a year.

Julia Bailey
Honorary Treasurer

NSPKU
The National Society for Phenylketonuria (United Kingdom) Limited

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NSPKU SPRING DRAW 2004 PRIZE WINNERS

Prize No.	Prize	Ticket No	Name	Location
1	£500	37723	Mrs P. Neal	Milton Keynes
2	X-BOX + 2 Games	6471	J .Newble	Weymouth
3	Personal CD Player	24500	R. Kerr	Letterkenny , Donegal
4	Blackpool Leisure Beach- Vouchers	17707	J Emerson	Bristol
5	£25 M & S Vouchers	34075	Moss	Crowborough
6	£25 Book Vouchers	24246	H Antonelli	East Lothian
7	£25 M & S Vouchers	14793	G Walker	Westhill, Aberdeenshire
8	£20 W.H.Smith Vouchers	12151	A. Rahim	
	Donated back to the Society-Thank you			
9	£20 H.M.V Vouchers	23900	A. Tordoff	Huddersfield
10	£20 Virgin Vouchers	10163	I Watson	Scunthorpe

Thank you to all who bought and/or sold Raffle Tickets on behalf of the Society , especially those who asked for more tickets .

The Prize for selling the most number of Raffle Tickets went to Helen Scally who with the assistance of her son Kian sold £282.50 of Raffle Tickets. Well done Helen & Kian !

The total raised was just over £5,706.64

Eric Lange

20th April 2004

NEWS & VIEWS 109/110
From Your Chair..

Recently I have once again had the pleasure of holding the stock of NSPKU merchandise. Taking on this work again has, I confess caused me some pleasure and some pain. A pleasure because it has already given me the opportunity to journey down to Taunton for the Bristol and Area Study Day – an event expertly organised by Diana Rawlinson, dietitian and with the skilled assistance of Dr Mike Webster and a myriad of others. David, Josh and I (Rosanna, was busy rehearsing for a show) had the opportunity of meeting a group of wonderful families, every one, like all of you reading this, dedicated to doing their best to provide for the PKU's in their midst.

I describe taking on the merchandise again as a pain solely because of the sheer volume of goods that we have for sale that need storing. You see, over the years our enthusiasm has outstripped your need, and despite our small-scale attempts at reducing the amount, I am still left with large piles and boxes skulking in the corner of the garage. What we have decided to do is to distribute the items that are not fast-selling, to clinics along with information about the Society, with the aim of increasing our membership. We want to continue to encourage as many PKUs, their families and carers as possible, to join. Whether or not they want to be active members we believe that being a member of the NSPKU brings many benefits.

What of our last Merchandising Officer, Sarah Taylor? Sarah took over the role from me and did an excellent job for many months until the pressures of her work increased and she was obliged to release the role in order to devote more time to her family and career. I would like to say a big thank you to Sarah who through her connection with us, having a good friend whose child has PKU, worked hard to help us and did a great job.

Please note, we have a few pairs of Salter scales left. They are a good kitchen model that weigh in 1g increments and normally retail at about £40. They have an add and weigh facility and are guaranteed directly by Salter, a company with whom we have experience of an excellent after-sales service. Cost is £28 with £4 P+P. Also the Kenwood Juicers are now priced at a very low £5 plus £4 P+P. Don't forget to send us your favourite drinks recipes so that we can share your ideas by printing them in News & Views!

Sara Bartlett
Chair

Donations

Donations to 31/10/04 **(£30.00 and over)**

Rosa Bartlett	56.00
Shirley Osborne	85.00
Ivan Vernon	75.00
Helen Scally	41.00
LD Thomas	50.00
The Queen's School, Wisbech	385.00
Cathy Darby	355.00
Christine Clothier	30.00
Three Legged Cross WI	300.00
Margaret Hokins (& Alison)	1,114.00
Mr Philip Squire	100.00
RAF Cosford	1,000.00
Grace Henderson	75.96
Liz Owen	60.00
Andrew Cardy (via Christine and John)	50.00
Merseyside Police	1,867.00
John Skidmore	1,012.00
Lesley Thomas	305.00
Norman & Pat Kerridge	1,004.83
Wendy Cheale	300.00
Pat Linton	3,803.00
Eleanor & Harry Gibbings	150.00
Neston Primary School	1,000.00
Capanac	242.42
Miss KM McCabe	40.00
John Skidmore	1012.00
Lesley Thomas	305.00
Mr & Mrs Kerridge	1004.83
Wendy Cheale	300.00
Pat Linton	3803.00
Mr & Mrs Gibbings	150.00
Gift Aided	
Sara & Dave Bartlett	133.23
DK Osborne	85.00
PD Rose	50.00
In memory of:	
Mrs J Aylett	395.00

Eh Up! Fundraising Fun in Yorkshire

Fine weather, after a week of wind and rain, set the scene for the 5th annual PKU day hosted by John & Sharon Skidmore in Normanton, West Yorkshire.

The seclusion of a private cul-de-sac provided the perfect setting for a congenial afternoon of fun, festivity and fundraising in aid of the NSPKU. The gods were kind and bestowed good weather for a perfect fun day.

Refreshments were provided for everyone, before they were encouraged into the "market place" to have their pockets, wallets and purses summarily emptied- all in a good cause. The method of emptying said pockets included:

Tombola stall
Raffle
Toiletries stall
Stationery
Plants
Flowers

Families of children with PKU from across the region attended and well over 130+ people attended throughout the day.

"Bob's your uncle" entertained the kids with a whole host of activities throughout the day, which was rounded off with a dancing finale involving most people who attended – young and old!

The raffle was drawn towards the end of the day, when the twelve lucky winners were announced. Donations received before, during and after the event helped the total amount raised reach a staggering £1,012.

The aim of the day is for families to come together in supporting each other, share experiences, have fun and raise funds for NSPKU. The event would not be possible without all the help and support from so many people, who so freely give their time to make this event such a huge success.

John Skidmore

Bobbies on Beat get Blisters on Feet!

Carl Beckley and his colleagues from the Merseyside Police Support Group gave up some of their valuable free time to raise a phenomenal £1,719.00 for the Society. Not content with pounding the Merseyside pavements, the policemen took on the arduous Lyke Walk.

The energetic coppers walked 40 miles across the North Yorkshire Moors, sustained by generous donations of food and drink from Asda and transported by the kindness of the Arnold Clark Van Rental Company.

Carl's 9 year old son Joe has PKU and must be very proud of his Dad's heroic endeavours. We at the NSPKU are extremely grateful and can assure Carl and his co-workers that their hard earned cash will be put to excellent use. We also hope their sore feet recovered in time for their next assignment!

Letters

News & Views, 51b The High Street, Watton-at-Stone, Hertford SG14 3SX
Email: editor@nspku.org

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,

Please find enclosed a cheque made payable to NSPKU for the amount of £1,000.

The money was raised at our annual "Year 6 Charity Fete", an event organised by our children in Years 5 and 6. The weather was very kind to us and the sun shone brightly. There were many stalls and we were very well supported by many local businesses who donated prizes. The head teacher was soaked in the stocks and one teacher was sponsored to dress as a fairy for the afternoon.

This year NSPKU was our chosen charity, as one of our pupils has PKU. We surpassed last year's total and I'm delighted to be able to forward the cheque to you.

The cheque was formally presented to Mrs. Alison Sheridan, who accepted it on behalf of NSPKU, at our leavers' service.

The photos enclosed show Mrs. Alison Sheridan and Emma Sheridan with the cheque.

Very best wishes to all concerned,

Rob Golding
Key Stage 2 Co-ordinator, Neston Primary School,
Cheshire.

Dear News and Views,

Please find enclosed a photograph of Andrew after abseiling down the side of the Maternity block at Ipswich Hospital on Sunday 18th July 2004, in aid of the NSPKU.

Also enclosed is a cheque for the sum of £50 which he raised through sponsorship. Andrew has recently passed his A levels and starts at Sheffield University in September, reading computer graphics.

We would like to take this opportunity to thank the Society for all the support we have had over the years.

Best wishes,

Christine and John Cardy.

Dear News and Views,

A friend of mine is a professional actress and offered to do a private performance of one of her monologues to raise money for NSPKU.

The play was "Bed Among the Lentils" by Alan Bennett about a vicar called Geoffrey and his wife Susan, who struggles to fit in with Geoffrey, the church and his eager army of ladies eager to help. This bittersweet comedy comes from the acclaimed series "Talking Heads" and has been performed on TV. The 40 min monologue was followed by live jazz whilst everyone enjoyed a drink and a chat after the performance.

It was a great evening which everyone enjoyed and we raised £300 for the NSPKU.

If anyone is interested in this type of entertainment, "Entrees" is the company name and it brings professional theatre to your home, to accompany pre dinner drinks, as a start to your fund raising event or conference or in a lunchtime recess. It provides light hearted entertainment to your event and provides an amusing alternative for your guests to enjoy.

Contact Ruth Redman on 01883 724502, email ruthredman_entrees@hotmail.com for more information or a brochure.

Best wishes,

Wendy Cheale
Mother of Alexander Cheale (4 years with PKU)

Dear News and Views,

Please find enclosed a cheque for £242.42 towards your fundraising.

This money was raised recently by "rattling a bucket" at a local night club and asking customers for donations.

I have to thank Ms. Jackie Granville for her efforts in this regard as she actually rattled the bucket personally! You will already have received two cheques over the last few months for £500 from Capanac Leisure. These were also the results of Jackie's efforts.

I wish you continued success with your fundraising efforts and I will continue to forward and monies we manage to raise.

Yours sincerely,
Paul Hanson
PKU Dad.

Letters

News & Views, 51b The High Street, Watton-at-Stone, Hertford SG14 3SX
Email: editor@nspku.org

Dear News and Views,

It's been quite a while since we've been able to do any fundraising for the NSPKU as we've been fostering and adopted a little boy we've had since he was three weeks old. He has spina bifida and hydrocephalus, is paraplegic and a wheelchair user. Caring for him is a different sort of challenge to a PKU diet.

Our own son was on the PKU diet (average of 3 exchanges a day) from 21 days to 21 years of age. He's now 27 and is doing OK. I could write a book about the trials and tribulations, fun and traumas we encountered, if only I had the time.

We have always been very grateful to the Society, News and Views, the conferences and the families we met at them for their encouragement, support and friendship we've had since joining in 1979/1980.

We've tried to give back a token of thanks for the Society's help in enabling our child to develop normally. To date, we think we have given £8,000 including the cheque enclosed for £1004.83.

We read with interest the articles in News and Views and hope our fundraising will help the NSPKU to carry on their good work. We know from our experience on the Council of Management many years ago that the funds are spent wisely.

It's been a pleasure to raise money and to make people aware of the condition. Nearly every parent knows about the Guthrie Test, but not many know what it is testing for. Thanks to the NSPKU, more people can become better informed.

A last note; we have been able to keep in touch with several families with children of a similar age to our own. Unfortunately we have lost the address of Sue Hacking and her PKU son Daniel who we think were from Nelson in Lancashire. It would be great if anyone could help us get in touch again.
Best wishes and thanks,

Norman and Pat Kerridge.

Editors note
If Sue, Daniel or anyone who knows their address would like to contact our administrator Lucy Welch, she will pass the details one.

Dear News and Views,

I am pleased to enclose a cheque for £175 as a donation to the NSPKU. One of our gorgeous little grandsons, Séamus (pictured with Santa in a recent issue!) has PKU and our daughter-in-law, Kiri, is sub-editor of News and Views.

On reading your last issue, I was impressed by the fundraising efforts of so many people and I decided to put my pastime of beading to good use. This summer I have been busy making bracelets and necklaces, whilst my daughter Elin obtained commissions from her colleagues at the BBC. This money is the profit made form the sales of these "jools"!

Looking forward to your next issue,

Lesley Thomas.

Dear News and Views,

Ciara Culligan and I, (both Dieticians at Norfolk & Norwich University hospital) together with Dr Thalange, Consultant Paediatrician & Dawn Moore, Dietetic Assistant, took a group of 8 Children & teenagers with PKU from the Norwich area bowling on 6th August 2004. We then provided a PKU lunch for them in the party room of the bowling alley (see photo attached – 8 out of 9 sitting have PKU).It was the first time they really met one another & many exchanged mobile numbers! Everyone seemed to enjoy themselves & I was very impressed by the standard of bowling! We had a quiz about PKU over lunch & Michelle Bernson & Jessica Whyard- Rowlands were joint winners receiving cinema tickets as their prize.

The other PKU kids there were Adam Scott, Steven Whyard, Sam Law, Charlotte Tink, Melissa Moss, Suzanne Osbourne.

Best wishes,

Rachel Pereira
Senior Paediatric Dietitian
Department of Nutrition & Dietetics
Norfolk & Norwich University Hospital
Colney Lane
Norwich
NR4 7UY
01603 287011

Dear News and Views,

Please find enclosed cheques totalling £1,114 as a donation to the NSPKU.

I raised this along with my cousin Alison doing the Women's Britannic assurance 10k run in Glasgow on the 16th May 2004. This is the second year I have done the run but it was Alison's first. I don't know if I can persuade her to do it next year!

My daughter Carys aged 5 has PKU and is pictured here with me (on the left) and Alison (on the right). I'm not sure who is holding who up!

I won't commit myself to next year just yet but as long as my legs will carry me I will have a go at raising funds for this excellent cause.

I would like to thank Alison, my colleagues at Weatherproofing Advisors Ltd., and my family and friends for helping me to raise this money.

Kind regards,

Margaret Hopkins.

Recent Advances in Cell and Gene Therapy for PKU

It has been a long time since National PKU News has reported on the progress of basic research aimed at correcting the defect in PKU at a molecular level. I asked Dr. Cary Harding to give us an update on recent research in this field. Dr. Harding is Assistant Professor of Pediatrics, Molecular and Medical Genetics as well as Medical Director of the Biochemical Genetics Laboratory at Oregon Health and Science University in Portland, Oregon. He has been involved in PKU research for 13 years. He describes his own exciting research as well as that of other laboratories

A permanent cure for PKU that would render dietary therapy obsolete is certainly the "Holy Grail" of many patients, their families, and their caregivers. Because of the outstanding success of dietary therapy, I often must defend my choice of PKU as a gene therapy target to medical professionals (and grant reviewers) who do not deal with PKU on a daily basis. However, in writing this article for families who live with PKU, I suspect that their desire for a permanent cure will be obvious and that I will need to provide little justification for gene therapy research directed at PKU.
How close are we to having a safe, effective, permanent cure for PKU? As I write this, there is no available gene transfer method that would fulfill these criteria. But recent work with the Pahenu2 mouse, the so-called "PKU mouse," has produced some promising results. In this article, I will review these recent experiments and the current status of cell-directed therapies for PKU. The focus will be upon two separate techniques: hepatocyte-mediated therapeutic liver repopulation (don't worry, I'll explain this later) and direct gene transfer using adeno-associated virus (AAV) vectors (I'll explain that, too).

> My dream for the future is to have an easy, safe, effective method for treating a newly diagnosed infant with PKU that will lead to permanent PAH production, normal blood phenylalanine levels, and will make diet therapy unnecessary.
>
> Dr Cary Harding

A Lesson In Physiology

First, a little biology lesson. This may be a simple review for some readers, but understanding the biology behind both normal phenylalanine (phe) metabolism and PKU is absolutely critical to understanding the goals and limitations of PKU gene therapy.

In the typical American diet, an adult ingests about 6,000-10,000 milligrams of phe per day as part of dietary protein. After digestion and absorption in the intestine, dietary phenylalanine travels to the liver. The liver is the central decision-making point for what happens to the phenylalanine. About 10% of it is needed for the synthesis of new proteins in the body. This amount is sent out into the bloodstream to supply all of the various organs with their daily phenylalanine supply. The remaining 90% must be disposed of. Normally, this occurs through conversion of phenylalanine to another amino acid, tyrosine. This chemical reaction is performed by phenylalanine hydroxylase (PAH), an enzyme produced in the liver specifically for the purpose of metabolizing phenylalanine.

In individuals with PKU, mutations in the Pah gene (the code the liver uses to make the PAH enzyme) prevent production of sufficient PAH enzyme. Excess phe accumulates in the body, in blood, and in every other tissue, to very high levels. PAH deficiency and elevated phenylalanine levels do not harm the liver, but phenylalanine does adversely affect the brain. Although the exact mechanism is still not understood, high blood phenylalanine causes poor brain growth and development and leads to small head size, seizures, and mental retardation.

The key to treating PKU is to reduce the amount of phenylalanine in the body. The PKU diet accomplishes this goal by reducing the intake of protein from food and therefore reducing phenylalanine in the diet. PKU researchers have proposed several different methods that do not rely upon dietary therapy to reduce phenylalanine levels.

Enzyme Supplementation Or Replacement Therapy

Previously in National PKU News, Dr. Christine Sarkissinian from Montreal, Canada has described her work with the yeast enzyme, phenylalanine ammonia lyase (PAL). She produced and purified the PAL enzyme in the laboratory and administered it orally to PKU mice. PAL worked in the intestine to break down some (but not all) of the phenylalanine in the mouse diet before it could be absorbed. BioMarin Pharma-ceutical, a biotechnology company in California, has been trying to further develop PAL for clinical trials in humans. Scientists at BioMarin are working to develop a form of PAL that can be given intravenously. In this approach, PAL would metabolize phenylalanine directly in the blood.
In a similar treatment approach, Dr. Raymond Stevens and his colleagues at the Scripps Institute in San Diego, California are developing a form of the human PAH enzyme that can be administered intravenously. Enzyme replacement therapy with

Continued on page... 10

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Promin Low Protein Pasta in Sauce Is a brand new fast food product available in two tasty flavours.

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Simply mix with water in a saucepan bring to the boil and then simmer for 10 minutes.

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Promin pasta in sauce is convenient and easy to cook. It provides an instant meal solution for lunchtimes and tea times, you could even serve it for dinner with a side salad.

Promin pasta in sauce will soon become a necessary addition to your kitchen cupboard. Available direct from Firstplay, see the contact details below.

Full analysis available on request.

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Promin Pasta and Promin Tri-Colour Pasta are now a well established brand and are available on prescription. Our pasta range also includes Imitation Rice, Pasta meal (available by prescription) and Lasagna Sheets (available by mail order). More recently interaction with PKU patients, parents and Dietitians has resulted in the expansion of the Promin range.

New products have been designed to reflect the needs of a maturing PKU population leading busy hectic lifestyles. Call us for more information or with your dietary suggestions.

Firstplay Dietary Foods produce low protein foods especially for your diet.

Please consult your dietitian about how to count these meals in your diet as they contain phenylalanine.

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human PAH is attractive because PAH is the native enzyme while PAL is a foreign protein. Therefore, the body's immune system would probably be less likely to reject PAH than PAL. However, unlike PAL, the PAH enzyme requires a vitamin cofactor called tetrahydrobiopterin (BH4). So enzyme replacement with PAH might also require administration of extra BH4 to keep the enzyme active. Enzyme replacement therapy with either PAL or PAH will likely require repeated infusions to be effective, but could result in a temporary decrease of blood phe levels and allow a less restrictive diet.

Therapeutic Liver Cell Repopulation

The central problem in PKU is the lack of PAH enzyme in the liver. Liver transplantation would permanently cure PKU since the donor liver would have the correct Pah gene sequence and would produce normal PAH enzyme. Liver transplantation is an acceptable form of therapy for several metabolic diseases in which dietary therapy has not been as successful as in PKU. However, a liver transplant is obviously a major surgery that carries a significant risk of death. Liver donors are difficult to find, and after liver transplantation the recipient of the new liver must take drugs to suppress the immune system and prevent their body from rejecting the liver. These immuno-suppressive drugs are expensive, must be taken for life, and reduced immunity may allow severe bacterial or viral infections to occur. Still, one PKU individual in Europe has reportedly undergone successful liver transplantation.

Rather than transplant the entire liver, several research groups have been exploring the transplantation of isolated liver cells as therapy for a variety of liver diseases. The major cell type in the liver and the cell that contains PAH activity is called the hepatocyte. The theory for PKU is that transplantation of even a few normal hepatocytes into the liver of an individual with PKU would provide enough PAH enzyme to lower the blood phenylalanine levels. This approach is sometimes called therapeutic liver repopulation because it repopulates the PAH deficient liver with new hepatocytes that do make PAH.

In my laboratory, we have been exploring therapeutic liver repopulation as a treatment for the PKU mouse. The big question before starting this research was: How many new hepatocytes will be required before the blood phenylalanine levels come down? Individuals with milder forms of PKU, so-called benign hyperphenylalaninemia, have blood phenylalanine levels that are only slightly elevated even on a normal diet and do not require dietary therapy. In these individuals, the liver makes 5-10% of the normal amount of PAH enzyme, but this small amount of PAH is present in every hepatocyte. In therapeutic liver repopulation, only the transplanted hepatocytes will make PAH enzyme. If we could replace 5% of the patient's hepatocytes with donor hepatocytes that produce the normal amount of PAH activity per cell, would this still be sufficient to lower the blood phenylalanine levels?

The First Experiments

In our initial experiment, we simply tried injecting normal hepatocytes directly into the blood vessels that feed the liver of the PKU mouse. Several weeks after this treatment, the blood phenylalanine levels had not changed and we could find no, or only very few, PAH producing hepatocytes in the liver. This turned out to be a simple mathematical problem; well, simple to calculate, but much harder to solve!

The adult mouse liver contains about 100 million hepatocytes (wow!); we injected about 500,000 hepatocytes. So we would have added only 0.5% new hepatocytes to the liver—not enough cells to cure the animal. Unfortunately, injecting more than about 500,000-1 million hepatocytes led to fatal blood clots in the animals.

The liver is one of the few organs in an adult that is capable of regeneration. That is, if the liver is damaged or a part is removed, the remaining liver cells will begin to divide and within days will have replaced the damaged or removed section with new cells. We reasoned that if we surgically removed part of the liver, then immediately transplanted in 500,000 cells, the new cells would grow and divide along with the native hepatocytes. This should yield greater numbers of PAH producing hepatocytes in the PKU mouse liver. Unfortunately, mathematics and physiology get in the way here too. The native and donor hepatocytes grow and divide at the same rate. So, if we take out half the liver (a procedure called a partial hepatectomy) leaving about 50,000,000 cells behind, then transplant in 500,000 new hepatocytes and let regeneration occur, each cell population will have doubled in size: to 100,000,000 for native cells and 1,000,000 donor hepatocytes. That is in fact what we found in our experiment: only about 1% PAH producing cells in the liver. The blood phenylalanine levels remained elevated.

The trick to making therapeutic liver repopulation work is to somehow give the donor hepatocytes a survival advantage over the native cells. That is, we need a method that allows the donor hepatocytes to divide but prevents the native cells from regenerating. If we could develop such a method, then the math becomes a bit more favorable. After partial hepatectomy and hepatocyte transplant, 500,000 transplanted hepatocytes will expand to 50,000,000 while the 50,000,000 native cells that remain will not have divided. We would achieve

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50% repopulation, more than enough to completely correct blood phenylalanine levels.

Studies With The FAH Mouse

To prove that hepatocyte-mediated therapeutic liver repopulation would work for PKU, we took advantage of a mouse with a different disease. The Fah?exon5 mouse, developed in the laboratory of Dr. Markus Grompe here at OHSU, has a disease called tyrosinemia type I. This is caused by deficiency of an enzyme called FAH that is further down the phenylalanine metabolic pathway. Humans and mice with tyrosinemia type I develop severe liver and kidney disease; without treatment, affected individuals die of liver failure. Fortunately, there is a new drug called NTBC which blocks the liver and kidney damage and prevents death in both humans and mice with FAH deficiency.

Dr. Grompe and his colleagues have demonstrated that normal hepatocytes transplanted into an FAH deficient mouse have a significant survival advantage over the native cells. Following the hepatocyte transplant, NTBC therapy is stopped. The native hepatocytes in the liver, lacking FAH enzyme, begin to die while the transplanted hepatocytes, which have FAH activity, rapidly divide to repopulate the liver. By four to six weeks after the transplant, over 90% of liver is repopulated with normal hepatocytes and the biochemical abnormalities associated with tyrosinemia are completely corrected.

To test hepatocyte transplantation in PKU, we actually bred the PKU mouse to mice with tyrosinemia in order to produce animals that have both diseases. In these mice, both phenylalanine and tyrosine are elevated in blood. The drug NTBC, which we add to their drinking water, is required to prevent liver failure. We have now performed hepatocyte transplantation in sixteen FAH-deficient PKU mice. NTBC therapy was stopped at the time of the transplant; blood phenylalanine levels began to decrease significantly at 30-40 days after the transplant as the extent of liver repopulation with normal hepatocytes increased (See figure, page 2). In examining the livers of these mice, we found a strong correlation between the amount of repopulation and the blood phenylalanine levels. Repopulation with as few as 2-3% normal hepatocytes caused the blood phenylalanine to drop from about 1800 µM (30 mg/dl) to about 1200 µM (20 mg/dl). With 5-10% repopulation, serum phenylalanine dropped to 300-600 µM (5-10 mg/dl). Blood phenylalanine levels were completely normal (< 200 µM) in animals that had more than 10% repopulation.

We drew two important conclusions from this experiment:

PKU can be cured with hepatocyte-mediated therapeutic liver repopulation if the donor cells have a selective growth advantage over the native cells. Correction of blood phenylalanine levels requires PAH activity in only a minority (5-10%) of the hepatocytes in the liver.

This is good news for gene therapy too, because we now know that we only need to produce PAH activity in about 10% of the liver hepatocytes to cure PKU. The major limitation in using hepatocyte transplantation to treat human PKU is the need to give the donor hepatocytes a selective advantage over the native cells. Several investigators, including myself, continue to work on this problem and are trying to develop an easy method of giving the donor hepatocytes a growth advantage without having to resort to a genetic trick like FAH deficiency. Hepatocyte transplantation has actually been attempted in about a dozen humans with liver diseases other than PKU. In most cases, without a survival advantage for the donor cells, the amount of repopulation was not enough to cure the disease. However, in two patients with diseases that cause liver damage, more extensive repopulation occurred and the signs and symptoms of the diseases did improve. Unfortunately, the effect was temporary (2-4 weeks) because the donor hepatocytes, being foreign cells, were eventually rejected by the patients and were destroyed by their immune systems. Another problem is the limited availability of donor livers, the same problem that limits the use of whole liver transplantation.

Will Stem Cells Be The Answer?

To overcome this difficulty, several investigators including Dr. Grompe and his team, are exploring the transplantation of bone marrow stem cells as a method of repopulating the liver. Stem cells are very special cells that have the potential of becoming just about any tissue type. There are very few true stem cells in the body —maybe only a dozen or so in the entire bone marrow of an adult. But there is hope that these cells can be obtained from a donor, transplanted into the liver of another individual, and coaxed into becoming healthy, normal hepatocytes. The future for therapeutic liver repopulation is quite optimistic, but much work remains before a truly viable treatment approach will be ready for clinical testing.

Gene Therapy For PKU

Gene therapy holds the promise of a permanent cure for PKU by restoring a fully functional Pah gene. Ever since the Pah gene was first isolated in the laboratory of Dr. Savio Woo in 1982, the goal of several investigators has been to find a safe, effective method to permanently transfer a normal copy of

Continued on page... 12

the gene into hepatocytes or other tissues and to restore phenylalanine metabolism. This research has been greatly facilitated by the availability of the PKU mouse for testing gene transfer methods. After many years of work, recent experiments in the PKU mouse have produced the most promising results yet. But enthusiasm for these recent successes is tempered by less than stellar results in the few gene transfer clinical trials that have been attempted in humans.

The primary site of phenylalanine metabolism is the liver, so the liver is the natural target for gene transfer. However, PAH enzyme is also produced naturally in kidney and pancreas; transfer of the Pah gene into these tissues or even tissues that don't normally contain PAH might lead to improved phenylalanine metabolism and lower blood phenylalanine levels.

In my laboratory, we have explored the possibility of producing PAH enzyme in skeletal muscle or in bone marrow in the PKU mouse, while the research teams of Dr. Beat Thöny in Switzerland and Dr. Thomas Jensen in Denmark have worked to produce PAH in skin cells. All of these approaches have been hampered by an inadequate supply of tetrahydrobiopterin (BH4, the vitamin required for PAH activity) in the target tissues. Work continues to overcome this obstacle, but to date, these approaches have not resulted in a permanent cure in the PKU mouse.

Inserting The Gene With Viruses

The most promising results have come from gene transfer experiments that target the liver. The liver naturally produces its own BH4, so its supply will not limit the effectiveness of gene therapy. Several different methods for transferring genes into liver cells exist, but the most successful attempts have used Nature's own method for injecting genes into cells: infection with a virus. Viruses naturally infect cells and carry in their own viral genes. After the virus penetrates into a cell, the virus uses the biochemical machinery of the host cell to make multiple copies of itself. In the final step, the newly formed virus particles break out of the host cell, sometimes killing the host cell in the process, and then continue on to infect other cells.

To use a virus as a vehicle for transferring therapeutic genes into cells, many of the normal viral genes are removed and replaced with the therapeutic gene. These altered viral particles are still capable of infecting cells, but because crucial virus genes have been removed, they cannot replicate themselves or produce new infectious virus particles.

Adenovirus is a highly infectious virus that causes the common cold. Adenovirus is capable of infecting almost all tissues of the body and is quite effective in penetrating liver cells. Adenovirus vectors that have been altered to carry therapeutic genes have been one of the most commonly employed liver-directed gene transfer methods. Researchers in Dr. Savio Woo's lab designed an adenovirus vector that carried the human Pah gene and injected this virus into the liver of the PKU mouse (Fang 1994). Liver PAH activity was restored in treated mice by 4 to 7 days after injection of the virus, but the amount of PAH activity varied between 5-20% of normal. Blood phe levels were completely corrected to normal in the animals that had more than 10% PAH activity. Unfortunately, blood phe began to slowly rise in all treated animals and had returned to pretreatment levels by 2 weeks after virus injection. Repeating the treatment had no effect on blood phenylalanine levels.

The immune system in mammals has evolved to quickly detect and destroy invading viruses. The treated PKU mice had developed a very strong immune response to the presence of the virus which destroyed the infected liver cells and eliminated the PAH enzyme. Following a second virus injection, the immune system immediately recognized the invading virus and eliminated it before it even had a chance to infect the liver.

Injection into the liver of a similar adenovirus vector has actually been used in a human clinical trial designed to treat ornithine transcarbamylase (OTC) deficiency, another inborn error of amino acid metabolism. That trial was stopped because of a similar problem with an immune reaction to the virus. Jesse Gelsinger, an 18 year-old man with OTC deficiency, received a large dose of an adenovirus vector that had been engineered to carry the OTC gene. Within hours of the infusion of the virus into the liver, signs of liver damage began to appear. All evidence pointed to a direct toxic effect of the virus. Jesse eventually lapsed into a coma and died of complete liver failure.

During the course of laboratory evaluation of adenovirus, a new type of virus was discovered. This previously unknown virus was found to be contaminating many adenovirus cultures that had been prepared from infected human tissues. This new virus is now called adeno-associated virus (AAV). Since the initial discovery of AAV, eight different subtypes of AAV, all differing slightly from each other and found in different mammals, have been described. AAV can infect many different tissue types but it cannot replicate itself unless an adenovirus has also infected the same cell. AAV by itself does not multiply within cells, kill the host cell,

Continued on page... 14

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or cause any known human disease. Probably because of that, the body's immune system does not mount a vigorous defense against AAV-infected cells. Administration of AAV vectors to animals has not in general caused significant inflammatory reactions like those seen with adenovirus vectors.

AAV also has the added beneficial ability of permanently inserting its genes into the chromosomes of the host cell. This property is useful for maintaining production of the therapeutic protein for long periods, perhaps even permanently. As in adenovirus vectors, most of the AAV genes can be removed and replaced with a therapeutic gene. AAV vectors have become a very popular method for transferring genes into liver.

Using AAV Viruses

My laboratory and the research team of Dr. Philip Laipis at the University of Florida have independently developed and tested AAV vectors for treating the PKU mouse. These viruses are from the second, or AAV2, subgroup. Our AAV2 virus and the virus produced by the Florida group differ somewhat in the exact DNA sequences that were engineered into the virus and this difference could affect the amount of PAH produced. In both cases, the virus was injected directly into the portal vein feeding the liver. About six weeks after injection, we detected a modest (about 30%) decrease in the blood phenylalanine levels of male mice only. On average, the blood phenylalanine levels decreased from about 1800 µM (30 mg/dl) to 1200 µM (20 mg/dl), a positive effect but certainly not a cure. Interestingly, blood phenylalanine levels did not drop much at all in female mice. Also, the effect lasted only about two weeks in the male mice before the blood phenylalanine levels went back up. Dr. Laipis' group had a similar but somewhat more successful experience (Laipis 2001). Blood phenylalanine levels dropped to below 600 µM (10 mg/dl) in male mice and were quite stable, lasting more than 40 weeks; but again, no effect was seen in female mice. The reason for this difference between males and females is not yet completely clear. The difference is probably related to hormonal effects. Liver in female mice appears to be less susceptible to AAV infection than liver in males. Also, blood phenylalanine levels tend to run higher in female mice than males so more PAH enzyme might be needed to lower the blood phenylalanine in female mice. These gender-specific differences have not been seen in humans.

Very recently, scientists in Japan, under the direction of Prof. Akihiro Kume, have reported the correction of blood phenylalanine levels of PKU mice treated with a new generation of AAV vector. This new AAV vector derived from the AAV5 subgroup appears to be more effective in delivering genes to liver than AAV2. Dr. Kume's group injected an AAV5 vector, at different but very large doses, into PKU mice via the portal vein (Mochizuki 2004). Within 2-4 weeks after injection at the highest doses used, blood phenylalanine levels completely normalized to about 1 mg/dl in male mice. This effect has persisted out to at least 40 weeks after infection. A familiar story was again seen in female mice. To fully correct the blood phenylalanine levels, a ten-fold higher dose of AAV5 was needed to treat female mice, and again, the effect did not last. The blood phe levels began to rise by 10 weeks after treatment and had risen back to the pretreatment level by 30 weeks.

Dr. Laipis' group has also developed an AAV5 vector for PKU. Their results were recently presented at the American Society of Gene Therapy meetings in Minneapolis, MN, and are very similar to those reported by Prof. Kume. Very high doses of AAV5 were required to lower the blood phe levels, with more virus needed for females than males. In male mice, blood phenylalanine levels were completely corrected and this effect lasted for at least 24 weeks after treatment. The effect was only temporary in treated female mice. Although AAV5 treatment of PKU mice did not appear to cause any symptoms of illness, there were potentially detrimental microscopic changes in the livers of treated mice. Long term monitoring of treated mice will be necessary to fully determine whether AAV5 therapy is harmless or not.

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The Italian Job

New Loprofin Lasagne sheets are just the 'Italian' job and are an addition to the authentic low protein Italian pasta range from SHS. Ideal for cannelloni as well as lasagne they do not need to be pre-soaked before cooking.

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A single human clinical trial of liver gene therapy using AAV has been performed. This trial was intended to examine the safety of AAV2 injection into liver of humans with hemophilia B, a blood clotting disorder. Low doses of AAV2 caused no harmful effects, but very little normal clotting factor (called Factor IX) was produced and no effect on bleeding tendency was seen. Therapeutic levels of Factor IX and a decrease in bleeding tendency was seen in an adult male that received a high dose of AAV2. Unfortunately, about four weeks after treatment, signs and symptoms of liver inflammation developed in this subject and the Factor IX in the blood rapidly disappeared. Though the researchers involved are still examining their data, it is thought that liver inflammation and loss of Factor IX production was probably due to an immune reaction against the Factor IX-producing liver cells. This is the first report of significant immune problems with AAV treatment; this immune reaction had never been seen in any animal experiment with AAV. For now, the clinical trial with AAV2 has been discontinued for safety reasons and until more is known about this episode.

Summary

Recent results with cell transplantation or liver-directed gene therapy in the Pahenu2 mouse have demonstrated the potential for a permanent cure of PKU, but these experiments have also uncovered significant biological problems that block the path to our goal. Obviously, many hurdles remain to be tackled before any human clinical trial can occur. The threshold for success is quite high. Strict compliance with the PKU diet will lead to normal intellectual capacity and a full healthy life for the person with PKU. Therefore, the safety of any cell trans-plantation or gene therapy approach for PKU must be rigorously demonstrated before widespread clinical use could occur. The potential benefits of any medical procedure need to outweigh the risks. Dietary therapy therefore remains the best treatment for PKU.

My dream for the future is to have an easy, safe, effective method for treating a newly diagnosed infant with PKU that will lead to permanent PAH production, normal blood phenylalanine levels, and will make dietary therapy unnecessary. I plan to continue working to that end.

Dr Cary Harding

Fate Special Foods recipes

FATE APPLE AND CINNAMON MUFFINS

Hello,
These muffins are perfect for any time of day. Have them for breakfast with a cup of coffee, pack them for your lunch, or have them for pudding after dinner. In this recipe, I use unsweetened pure apple juice, the kind you get in a carton, this will give the muffins a lovely flavour and moist texture. They are delicious hot or cold.

Ingredients

200g apples, such as Cox's, Braeburn or Granny Smiths, peeled, cored and chopped into small pieces.
300g Fate Low Protein All-Purpose Mix
150g soft light brown sugar
1tsp baking powder
1½ - 2 tsp cinnamon
120g soft margarine
240mls unsweetened pure apple juice

Method

Pre ~ heat the oven Gas 6 200°C 400°F.

Place the Fate All-Purpose mix into a mixing bowl. Add the light brown sugar, baking powder, and cinnamon. Stir.
Add the margarine and the apple juice.

Using a wire whisk, mix quickly for about a minute until well blended and smooth.

Stir in the chopped apples.

Divide the mixture between 12 large muffin paper cases set inside a muffin tin.
Bake in a pre ~ heated oven for 25 – 30 mins until risen and golden brown.
Place on a cooling rack to cool.

The muffins freeze well

Best wishes
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FATE SPECIAL FOODS ~ SPECIAL FOODS FOR SPECIAL PEOPLE

London says..... Yes We Cook That.com!

In February this year patients from the 3 London metabolic centres came together to taste a selection of foods made by Yes We Cook That.com.

Yes We Cook That.com is a company based in Glasgow, which makes a selection of low protein ready-made meals, and delivers these around the country. Patients with PKU and their families know only too well that there is no microwave, frozen ready made, or convenience low protein foods. So a chance to try these delights was very welcomed.

Great Ormond Street Hospital, Guy's Hospital and The National Hospital for Neurology joined forces and invited their patients with PKU or other disorders requiring a low protein diet. Guy's Hospital hosted the day in their catering suite on the 29th floor. So not only did everyone get great food to taste but also got a view of London which beats that of the London Eye!

Patients and their families got to know how to order the food and of course the chance to taste the delights of Mushroom and Sausage casserole, Roast Vegetable Lasagne, Roasted onion bread, shortbread biscuits, coffee cake and Barbie or Beckham birthday cakes plus lots more…

Dietitians from each hospital, a nurse specialist and a dietetic assistant came to help, but with over 100 people attending it was a bit of a challenge to keep the day running smoothly: - helping Pauline and Leanora get the food ready, whilst keeping an eye on any inquisitive toddlers and of course getting a chance to try the food out!

Everyone thought it was a success, the food vanished and food orders were made. Patients and their families also got a chance to mix with others from different hospitals – lots of telephone numbers and e-mail addresses were exchanged!

We'd like to say a big thank you to all who helped on the day, to the London branch of the NSPKU for their kind sponsorship and to Leanora and Pauline from Yes We Cook That.com for coming down from Glasgow and letting us taste such great low protein foods - hope you got lots of food orders!

Joanna Eardley
Metabolic Dietitian, Guy's Hospital, London

Dietitian's Report

EDALE 2005

The date for the Edale outward bound weekend is July 29th to 31st. The weekend is open to any child with PKU aged between 9 and 13 years. If you would like more information about the weekend or would like to make a provisional booking for your child please contact me. See inside front cover for my details.

AMINO ACID ANALYSIS RESULTS

You will all probably be aware that each year we try to analyse around 10 foods for their phenylalanine content. The results are through for the last analysis and these foods can be included in the PKU diet in the following way:-

Foods which can be counted as FREE

Blue Dragon Rice Flour Pancakes
Walkers Crackers – Cantonese Black Bean Flavour
Walkers Crackers – Thai Lemon Grass Flavour
Karela
Pak Choi

The following foods need not be counted in the diet but keep to the amounts given.

Soy Sauce – any Brand name or variety (i.e. light or dark) up to 2 tablespoonsful free
Tinned Sweet Chestnuts. Up to 100gms. Free.

Vegetable Crisps
Two types of Vegetable Crisps were also analysed. We chose crisps made from beetroot, parsnip, sweet potato (not potato or maize). The medical advisory panel has decided to have a few more of these types of crisps analysed so that we can give an exchange value for them all and not just one brand name. In the meantime take the protein value on the pack to calculate the amount for one exchange. If you are unsure about how to make this calculation ask your dietitian to explain this to you or to work it out for you.

A little note about some of these new foods which you may be trying for the first time.

Blue Dragon Rice Flour Pancakes.

You will find these in the Oriental section of the supermarket. The pancakes come in a thin cardboard packet and are about the size of a tea plate. They are very hard and thin. To prepare them you just slip one into a bowl of very hot water and leave for a few seconds. Fish the pancake out with a slotted fish slice or draining spoon and put onto a tea towel. The pancake will now be quite rubbery. You can then fill it with finely sliced and fried free vegetables such as a few beansprouts, water chestnuts, peppers, onion, mushroom and fresh spring onion. Add a spoonful of one of the free Blue Dragon sauces in the diet booklet and roll up. Serve with low protein rice and a little soy sauce, maybe some of the new free Walkers crackers (see above) or Sharwoods Ready to Serve Prawn Crackers and some salad and you have a super Chinese meal!

Karela
This is an Indian vegetable which looks a bit like a very knobbly small cucumber. It has quite a bitter flavour and is used in curries or served as a vegetable on its own. It is also available in tins.

Pak Choi
This looks like a small Cos lettuce or Chinese leaves. It is used in stir fries, can be braised in vegetable stock with a little grated or chopped fresh ginger or steamed. It can also be eaten raw and is a bit tougher than lettuce.

Tinned Sweet Chestnuts
These can be chopped and mixed with sauté onions, mushrooms and low protein breadcrumbs (or any other free food you fancy) and then use to stuff peppers, large mushrooms or to make a savoury vegetable rissole.

Other useful foods in the supermarket
Barbara Cochrane, dietitian in Glasgow, told me about two useful foods which also might go well with some of the foods above.

Amoy – Straight to Wok Rice Noodles
1.6g protein per 100gms.
65gms for 1 exchange

Uncle Ben's Aromatic Cantonese Sauce
Sweet Soy and ginger Sauce 0.6gms protein for 100gms.
This can be taken freely.

Incorrect Nutrition Information on Brand Foods
Kings Food seen in Morrisons
8 Hot Dog Sausages in brine have been labelled as containing no protein.
This is obviously wrong and the company have been alerted.

Dietitian's Report

Asda 10 Dark Choc Ices.
On the packet the label reads that each choc ice contains 0.3gms protein. This is incorrect. Each choc ice provides one exchange.

If you see a food with a protein value which sounds too good to be true, it is probably wrong. Give me a ring or ask your dietitian to contact me and I can get it checked out. Much better to be safe than sorry.

Robinsons Fruit Shoot Drinks
You may remember that Robinson's changed the way they distinguished the sugar containing drinks from those which are sweetened with aspartame. The sugar containing ones had a green bottle top which made it easy to recognise. They then changed this easy system and without confusing you all with what they changed to I can tell you that the old system is back again!

So, the drinks which are fine for the PKU diet and do not contain aspartame have a green top again! Hooray!!

Teething Gels
Diana Rawlinson, dietitian in Bristol, gave me some information about teething gels. The pharmacy at her hospital investigated these and the following are free from aspartame:

Bonjela Teething Gel
Dentonox Teething Gel
Anbesol Teething Gel
Nelsons Teetha Granules
Ashton and Parsons Infant Powders

Thank you Diana!

Eleanor Weetch
Society Dietitian

Birthday Celebrations for Bryan Pearce

Bryan Pearce is the foremost primitive painter in the UK. He has PKU and was born in 1929 before screening started. Despite the damage that he suffered through lack of treatment he found peace and fulfilment through his painting.

Anyone who uses email knows that when you switch it on the messages seem to fall into one of 3 categories. Despite virus checks there is junk mail to be deleted unopened; there is the work stuff which requires some thought and digging around for information and lastly and most pleasant of all are the messages containing some interesting news or an invitation to something. The message I received from Janet Axten, a trustee of Bryan Pearce, without question fell into the third category!

The message was that on July 21st it was Bryan's 75th birthday and to celebrate this event and honour him for his artistic talent Tate Britain on Millbank in London would be displaying one of his paintings in the entrance. My response was that I would definitely go to London to see the painting (take the children along too for a bit of culture) because I have only ever seen postcards of Bryan's paintings, never an original which are now worth a lot of money.

The next thing that happened was an invitation in the post to a tea party to be held at The Penwith Gallery in St Ives on July 24th. This actually coincided with a few days' holiday I was having with my family in Devon so it wasn't so far to go for a party! Now my geography, especially of the south compares with those southerners whose knowledge of the north stops at Watford. It's not good and I thought St Ives was a stone's throw from Barnstaple. A four hour journey by train was lovely. Beautiful scenery and most people on a Saturday, off on holiday, made a relaxed atmosphere.

The station at St Ives is tiny and of course doesn't go on anywhere else- we had reached the sea! Having arrived in good time for the 4pm tea I decided to go to visit The Tate art gallery in St Ives where I felt sure some of Bryan's paintings would hang. Actually there was only one there of the church. I bought every postcard in the shop reproducing Bryan's paintings and made my way to the party.

Bryan was seated with his carers beside him to welcome guests. "No presents" was the request so everyone had taken a card. There was quite a queue forming to say hello and give a card and within half an hour the gallery was filled with 250 people. I asked someone to tell me where I could find Janet and also Sir Alan Bowness, another trustee of Bryan's with whom I have had some correspondence. Both Janet and Sir Alan have been very helpful to the Society and encouraged me to publicise Bryan's wonderful story. Apart form these two people I knew no-one. I'm not an arty person so didn't want to get involved in any technical conversation about art so I decided to approach people with the opening line" and what is your connection with Bryan?" This brought about a surprising range of responses. One lady, struggling as I was with teacup and plate of sandwiches was really interesting. She had taught Bryan to play the glockenspiel (a musical instrument of bells or bars struck by hammers). Sadly Bryan had to stop because of increasing deafness but she said she made tapes for him of the notes, recorded in slow time which as he learnt she speeded up until he could do it at the correct pace. Apparently Bryan used to play "Drink to me only with thine eyes" until he drove everyone crazy!

Bryan cuts his birthday cake

Another couple, enjoying the Devon scones with cream on top, used to live in a flat close to Bryan and his parents at Piazza. Their children, now grown up, used to visit the Pearces who were obviously very sociable and kind and welcomed children into their house. Much to this couples' dismay they supplied those sweet necklaces so loved by little children but no-one else! The couple's brother had bought paintings of Bryan's 40 years' ago, a sound investment as his paintings now sell for many thousands of pounds.

Bryan Pearce
Interior, St. IA Church

At any birthday tea party there must be a cake. Bryan sat beside the cake, a trumpet was sounded and silence fell. Sir Alan Bowness spoke of Bryan's achievements and wished him a happy birthday. Janet then followed by first of all offering a special thank you to a number of people. I felt very proud to be the first mentioned for the NSPKU. I thanked Janet for this and said I was really delighted to be there. It was interesting then to be asked by several people "What exactly is PKU?" They had heard of it but I suppose like many things if it doesn't affect

Bryan reads his birthday cards

you or your family directly the interest isn't there to find out more, and it is complicated. What I was so pleased about was that everyone in the room now knew about the NSPKU.

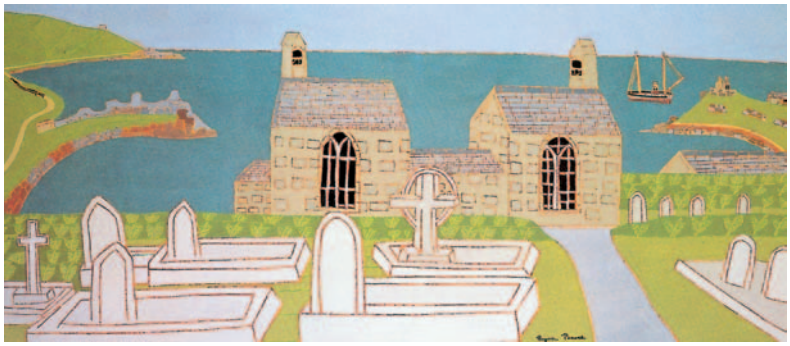
After the speeches, Clare, one of Bryan's carers who has looked after him since his mother died 6 years' ago helped him to his feet and prompted him to say a thank you to us all for coming and hoped we had a nice time. We sang "For he's a jolly good fellow" and the cake was cut.

I had to take a taxi to the station at St Erth; the last train out of St Ives leaves at 4pm. I started talking to the taxi driver who knew Bryan well as he often took him to different places. Recently he had taken him to the optometrist at the top of the town and when he was there he was asked if he would like to paint from that elevated point. Bryan said he would and that was organised for him. Bryan still paints 6 days a week.

Travelling back on the train I thought quite a lot about the day and Bryan. Yes, it was a tragedy that he was born with PKU before screening but for Bryan he has lived and still does live a rewarding and happy life surrounded by people who love and respect him and that must be something to be celebrated.

Unfortunately there are those who are severely affected by untreated PKU but who may still be helped by diet. The untreated PKU trial, we hope, will confirm this.

Eleanor Weetch
Society Dietitian



Bryan Pearce
The Cemetery, St. Ives (Tate Gallery Collection)

SHS recipes

Chilli & Pepper Pasta

Ingredients

- 75g (3oz) Loprofin Low Protein Penne
- 1 x 15mlsp (1tbsp) cooking oil
- 1 x 5mlsp (1tsp) salt
- 75g (3oz) red pepper
- 75g (3oz) yellow pepper
- 75g (3oz) green pepper
- 1 red chilli (approx 25g/1oz)
- 2 cloves garlic (skin on)
- 2 plum tomatoes (100g/4oz flesh)
- 6-7 fresh basil leaves
- 3 x 15mlsp (3tbsp) olive oil
- Salt and pepper
- Extra basil leaves for garnish

Method

1. Three quarters fill a large saucepan with water, bring to the boil, add the pasta, oil and salt to the pan. Return to the boil, stirring. Reduce the heat slightly and cook for 8 minutes. Drain thoroughly.
2. Meanwhile, place the peppers skin side up, on a baking sheet, cutting if necessary to ensure they lie flat
3. Deseed the chilli and place skin side up on the baking sheet with the peppers, Halve the garlic cloves and place skin side up on the baking sheet. Place the baking sheet under a preheated grill for a few minutes until the skins have blackened.
4. Transfer the peppers, chilli and garlic to a polythene bag, to sweat for a few minutes.
5. Halve and deseed the tomatoes and place under a preheated grill for 2-3 minutes until the skins shrink and can be removed easily.
6. Remove the skins from the peppers, chilli, garlic and tomatoes, process briefly or mash the vegetables, with the basil leaves and olive oil, to give a chunky sauce.
7. Transfer the mixture to a saucepan, add the cooked pasta, place over a moderate to low heat for approximately 5 minutes, until bubbling and heated through. Serve immediately, if desired garnish with fresh basil leaves.

Serves 1

Tuscan Style Pasta Stew

Ingredients

- 50g (2oz) carrot, thinly sliced
- 65g (2 ½ oz) onion, chopped
- 100g (4oz) celery, thinly sliced
- 125g (5oz) fennel, thinly sliced
- 2 medium cloves garlic, crushed
- 500ml (18floz) homemade vegetable stock
- 400g can chopped tomatoes
- 75g (3oz) **Loprofin** Low Protein Penne Pasta
- 2 x 5mlsp (2tsp) dried parsley
- 1 x 5mlsp (1tsp) dried oregano
- 1.25mlsp (½ tsp) salt
- 2.5mlsp (½ tsp) paprika pepper

Method

1. Place the prepared vegetables, garlic, stock and tomatoes in a medium sized saucepan. Bring to the boil, reduce the heat, cover and simmer for 10 minutes.
2. Stir the **Loprofin** Penne Pasta, herbs and seasonings, into the pan, bring to the boil. Reduce the heat cover and simmer for a further 10 minutes, stirring occasionally during cooking.
3. Serve at once with Low Protein Bread Rolls.

Serves 2

Tip:

If you have the time to chop fresh parsley, use 4 x 5mlsp (4tsp) in place of the 2 x 5mlsp (2tsp) dried, as this will improve the flavour of the dish.

At Birmingham Children's Hospital.....

Spooky, kooky, creepy, and fun! Goblins, ghosts, witches and vampires! These were the words that can only describe some of the scary happenings at our latest PKU school. For over 10 years, the Birmingham Children's Hospital PKU team has organized events for our children with PKU, so it was time to do something a little bit different. So what better than to have an 'eerie' event for our latest two PKU schools held in the October half term holiday!

With the awesome help of Gemma from SHS, Katie and Sue from Vitaflo, combined with our very own PKU team (Di, Anne, Louise, Pat, Nilusha and myself); we set about entertaining 25 of our 4-10 year old children in our Witches Den! We always teach plenty about diet and PKU but more importantly we wanted the children to have a fantastic time- it was in their holiday time after all! The children did a variety of things: they made low protein Witches fingers; monster pizza faces; created their own Vitabite spiders; had great fun 'apple bobbing'; 'Trick and Treating'; cutting out scary masks; made 'lolly-stick' ghosts, they sang Di's latest PKU single (Di is, of course, Birmingham Children's Hospital answer to Kylie!). They also produced paper mache PKU pumpkins and even devised their own gruesome treat for our 2 unsuspecting doctors. In fact, the days went so well we thought we would share a few of our simple ideas and photo shots with you. We might even share one or two of the children's jokes!

Scary Games

Paper masks
With just paper plates, coloured pens and paints, the children made their own PKU scary masks. They made anything from pumpkin faces, vampires, black cats, to ghost masks. These were then decorated with pipe cleaners, fluffy pom pom balls, home-made blood (artificial of course), tissue paper and food cut outs. Needless to say, by the time the children had finished they looked really frightening!

Pasta skeletons
Simply by substituting ordinary pasta for a variety of low protein pasta shapes, some of the children made their own pasta skeletons by sticking pasta onto black card.

Mummy Game Wrap
Using loads of white, cheap toilet paper, we wrapped each child up like an Egyptian mummy. Then to look really scary and authentic, we put drops of blood down the front of the body! The grizzlier it looked the better! The children loved it!

Low protein cooking: (thank you to SHS)

Witches Fingers

2 packets Loprofin choc chip cookies
100g (4oz) caster sugar
100g (4oz) syrup
50g (2oz) economy butter
50g (2oz) raisins, chopped

For decorating:
Icing sugar
Green food colouring
Black writing icing
Raisins

15cm (6inch) square tin

1. Combine the sugar, syrup and butter in a pan, place over a moderate heat and stir until melted.
2. Place the cookies in a strong polythene bag and crunch the biscuits finely, using a rolling pin.
3. Add the crushed cookies and raisins to the melted mixture and stir well, until thoroughly coated.
4. Transfer the mixture to a 15cm (6inch) square tin. Using a fork, press down well and level the surface.
5. Chill for 2 hours before cutting into long thin slices (or witches fingers!!!).

For witches fingers cover each 'finger' in green icing, use raisins for warts and black writing icing to create fingernail, hairs and wrinkles!

Makes 12 fingers

A cauldron full of "witch's brew."

Eye of lizard, toe of frog,
Tail of rat and bark of dog,
Sneeze of chicken, cough of bat,
Lick of weasel, smell of cat

Stir it up and mix it well
To make a magic
Halloween spell

This is one especially developed as a treat for the PKU doctors. A messy game but lots of fun!

We found a huge Witches cauldron (a large black bucket would have done). We asked the children to wrap up tasty treats for the doctors. These treats were placed at the bottom of the cauldron e.g.

sweets, fruit etc. Then the children filled up the pot with slimy and sloppy suitable foods for PKU (free and exchange foods) e.g. wobbly low protein jelly, tinned spaghetti, banana skins and apple cores, mashed potato squashed chips, and pickled onions. We bought some cheap fake eyeballs and maggots (and any other suitable delicacies). We asked the children to make up their own Halloween spell as they mixed it all together with a large stick.
The children loved it (…..and so did the dietitians) when the doctors had to dig deep in this hand-made 'slime bucket' for their hidden treats. What a mess!

Other Halloween things we made:

Large black cardboard bats which dangled from the ceiling
Ghost suckers: taking a round-headed lollipop, we placed the head of the lolly in the centre of the white tissue. Gathering the tissue round the head, it was tied tightly just below the head with a rubber band or string. We used black felt tip pen for eyes.
We stuck a paper face (e.g. witches or vampires) onto a flat lolly. The lolly stick formed the body and a black cloak was made from a black bin bag.
We made home- made pumpkins from paper maché (modelled round balloons), and then filled the inside with miniature play dough food displays.
The children wrote halloween PKU stories

And now for a joke…….

Ghost Sucker
Why did Dracula go to jail?

He tried to rob the blood bank.

The Spooks Step out

Halloween Story by Hattie Beech Aged 8 years

Once on a Halloween night, four spooks climbed into a carriage of a ghost train. They then stepped off the train into the fun fair. There was a skeleton, a ghost, a lady whose head fell off and a devil. They all liked fruit and had PKU. They went onto a very fast roundabout and felt very dizzy. The skeleton got them some candyfloss that made them feel better. Then the lady whose head fell off said 'Look a dodgem ride lets go!' But the skeletons bones fell apart. "Lets fix you" said the devil. Once they had fixed him they hopped quickly back into their places and fell asleep. The next day everyone who went on the ghost train was very disappointed. The ghost train was closed down and this is why:

GHOSTS AND GHOULS ASLEEP. COME BACK TOMORROW!

FUN FAIR RIDES

NEW

Fasta Pasta

Available on Prescription for Phenyketonuria

at lasta...

Ideal for modern day living the unique new Loprofin low protein Snack Pot from SHS is a quick and simple to use convenience food.

Just add boiling water and you have savoury curry flavour pasta in only a few minutes, ideal as a snack or as a part of a main meal.

... low protein snack pot

Each 47g pot contains: Energy 694kJ, 165kcal, Protein 0.9g of which phenylalanine 23mg (½ an exchange per pot).

For more information on the Loprofin Snack Pot, please contact our Nutrition Services Department on 0151 228 1992.

SHS

SHS International Ltd, 100 Wavertree Boulevard, Liverpool L7 9PT, United Kingdom. Tel: +44 (0) 151 228 8161 Fax: +44 (0) 151 228 2650 Internet: www.shsweb.co.uk

NSPKU 32ND Annual Family Conference Weekend 2005

The Rendezvous Hotel, Skipton North Yorkshire

Friday 25th to Sunday 27th February 2005

NSPKU
The National Society for Phenylketonuria (United Kingdom) Limited

HOTEL REF - NSPKU 2005

Name (Please enter first and last names)	PKU Yes or No?	Age at 25.02.05 (if under 18 yrs)	Cot Needed? Yes or No?	High Chair Needed? Yes or No?	Childcare Needed? (Creche for 6 yrs & under) Yes or No?	Room Type Needed F=Family S=Single D=Double T = Twin	FEE TO PAY
Day Visitors – Days Attending (Please tick)			Saturday ☐		Sunday ☐		
TOTAL - £							

Card payments: If you wish to pay by credit or debit card please tick the appropriate box -:

SWITCH ☐ MasterCard ☐ AMERICAN EXPRESS ☐ VISA ☐ CARD NUMBER: ☐☐☐☐☐☐☐☐☐☐☐☐☐☐☐☐

Expiry date:..... Name of Cardholder:.....

Signature:.....

Deposit Payments: Adults (per person – 18 yrs+).....£30

Children (per child – 3-17 yrs).....£20 Total Deposit Payable: £_____

I enclose Cheque/PO (Do not send cash) to Rendezvous Hotel for: £_____

(Insurance cover is NOT included. Delegates should make their own arrangements)

*Full balance of monies should be paid to - "The Rendezvous Hotel" prior to arrival date.

(The due date will be confirmed to you by the Hotel)

ANY OTHER REQUIREMENTS :e.g. specific room request, wheelchair access, special diets (excluding PKU) etc

.....

.....

CHILDCARE FACILITIES: Number of children – 0-1 years ☐ 2-3 years ☐ 4-6 years ☐

CHILDREN'S TRIPS: (Enter Numbers) - 7 + years ☐

I AM ABLE TO ASSIST WITH SUPERVISION ON THE TRIPS: *(Please tick) ☐

Note:*Childrens' trips take place subject to adequate numbers of volunteer supervisors coming forward.

If this is the first time you have attended a Conference, please tick the box ☐

Signature:..... Date:.....

Contact Address:.....

.....Contact telephone..... E-mail:.....

Please send completed form and deposit payment to:
NSPKU 2005, The Rendezvous Hotel, Keighley Road, Skipton, North Yorkshire BD23 2TA.
Telephone: +44 (01756) 700100 Facsimile: +44 (01756) 700107
web site: www.rendezvous-skipton.com

Data Protection Act: The information given here will be used solely to communicate with you. It will not be divulged to any other person or organisation.

NSPKU 32ND Annual Family Conference Weekend 2005

The Rendezvous Hotel, Skipton North Yorkshire

Friday 25th to Sunday 27th February 2005

NSPKU
The National Society for Phenylketonuria (United Kingdom) Limited

Venue:
This years NSPKU Annual family conference weekend will be held in Skipton, North Yorkshire. This hotel provides us with an absolutely delightful venue to hold our annual 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 creche, 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:
75 en-suite bedrooms, training centre, 12 meeting rooms, the Mallard bar, H2O brasserie, 120 car parking spaces, playzone nursery, indoor swimming pool, whirlpool spa, steam room & sauna, solarium, gymnasium, 2 squash courts and hair & beauty salon.

(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 the PKU amino acid supplements and 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.

Around and About:

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

Rail: Skipton station – 1 ½ 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.

NSPKU 2005 Residential Conference Fees:

Non-PKU Adult (18 years +)	= £155.00
Non-PKU Child (under 18 years sharing with parents)	= £65.00
Non-PKU Child (under 18 years in own room/sharing with another child)	= £75.00
Adult single room weekend supplement (18 years +)	= £20.00
PKU Adult (18 years +)	= £77.50
PKU Child (under 18 years sharing with parents)	= Free
PKU Child (under 18 years in own room/sharing with another child)	= Free
All Infants (0-2 years)	= Free

Day Visitor Fees:

Non-PKU Adult & Children (coffee, luncheon & dinner)	= £15.00
PKU Adult & Children (coffee, luncheon & dinner)	= Free

Booking Conditions:

All conference bookings and fees MUST be made by (Monday) 31st January 2005.

Your NSPKU 2005 Conference Organiser is: Mike Bailey

Grove Fold, Claughton-on-Brock, Garstang, Preston, Lancashire PR3 0PU

Email: conference@nspku.org • Home Telephone: (01995)641119 • Website: www.nspku.org

NSPKU 32nd Annual Family Conference Weekend 2005

Programme of Events Fri 25th Feb

TIMINGS:	PROGRAMME CONTENT:	LOCATION:
1230 - 1930 Hours	NSPKU 32nd Annual Family Conference 2005 - Welcome Registration Desk Officially Opens in the hotel:	Main Reception
1700 - 1800 Hours	Civic Drinks Reception: **(NSPKU 32nd Annual Conference 2005 will be greeted by the Lord Mayor & Mayoress of Skipton, North Yorkshire)**	Malham & Winterburn
1815 - 1945 Hours	NSPKU 32nd Annual Family Conference 2005 Welcome Dinner in the hotel:	Malham & Winterburn
1945 - 2400 Hours	Evening Entertainment: NSPKU Spring Draw 2005 & **(DJ & "Pop Idol" Discotheque: Dress up as your favourite pop idol and win a prize!**	Malham

Meeting NSPKU Council of Management: (Conference Centre at 9.00pm)

Programme of Events Sat 26th Feb

TIMINGS:	PROGRAMME CONTENT:	LOCATION:
0700 - 0845 Hours	Full Breakfast will be served in the Hotel:	Malham & Winterburn
0830 - 1230 Hours	Professional Nursery/Playgroup: (Infants & Children Under 8 years of age)	Kildwick
	Children's Outing (to be advised): (Children over 8 years of age will be taken out this morning accompanied by adult carers)	Conference Centre
0900 - 0915 Hours	Opening Introduction - NSPKU 32nd Annual Family Conference (Topic: NSPKU General Introduction & Functionality)	Wharfedale Suite
0915 - 1000 Hours	Morning Sessions: Dr Shelly Channon (Topic: The Neuropsychometric outcome of adults with PKU on and off diet)	Wharfedale Suite
1000 - 1045 Hours	Jane Gick / Dr Neil Dalton (Topic: Taking blood samples)	Wharfedale Suite
1045 - 1115 Hours	Morning coffee/tea break & biscuits: PKU Food manufacturers exhibition stands will be open:	Mallard Bar Mallard Bar
1115 -1200 Hours	Dr Maureen Cleary: (Topic: Research and developments in PKU)	Wharfedale Suite
1200 - 1230 Hours	Feedback Session: (Topic: Morning session)	Wharfedal Suite

Programme of Events Sat 26th Feb

TIMINGS:	PROGRAMME CONTENT:	LOCATION:
1245 - 1400 Hours	Luncheon will be served in the hotel:	Malham & Winterburn
1400 - 1700 Hours	Free Time:	
1700 - 1900 Hours	"Children's PKU Party" : 11 years & Under Infants/Children PKU/NON-PKU all very welcome: (Entertainment: Suprise Children's Entertainer)	Kildwick
1730 - 1830 Hours	ANNUAL GENERAL MEETING:	Wharfedale Suite
1915 - 2100 Hours	NSPKU 32nd Annua; Family Conference 2005 Final Dinner in the hotel:	Malham & Winterburn
2100 - 2400 Hours	Evening Entertainment: (to be advised)	

Programme of Events Sun 27th Feb

TIMINGS:	PROGRAMME CONTENT:	LOCATION:
0700 - 0845 Hourss	Ful Breakfast will be served in the hotel:	Malham & Winterburn
0830 - 1230 Hours	Professional Nursery/Playgroup: (Infancts & Children under 8 years of age)	Kildwick
	Children's Outing (to be advised): (Children over 8 years of age will be taken out this morning accompanied by adult carers)	Conference Centre
0900 - 0945 Hours	Morning Sessions: Claire Rowlands/Anna Brazier (Topic: The clinical psychologist and me)	Wharfedale Suite
0945 - 1045 Hours	PKU Workshop/s: (2 x PKU Topic Matters will be covered) Workshop 1: (to be advised) Workshop 2: (to be advised)	
1045 - 100 Hours	PKU Wokrshop/s Feedback Session: - Wharfedal Suite	Wharfedale Suite
1100 - 1130 Hours	Morning coffee/tea break & biscuits	Mallard Bar
	PKU Food Manufacturers exhibition stands will be open:	Mallad bar

Programme of Events Sun 28th Feb

TIMINGS:	PROGRAMME CONTENT:	LOCATION:
1130 - 1230 Hours	"New for 2005" Nick Nairn? (Tpoic: Ready Steady PKU Cook!!!)	Wharfedale Suite
1230 - 1300 Hours	Eleanor Weetch/Eileen Green/Richard Ward (Topic: The basic kitchen. What's useful and what's not?)	Wharfedale Suite
1300 - 1415 Hours	Luncheon will be served in the hotel:	Malham & Winterburn
END	NSPKU 32nd ANNUAL FAMILY CONFERENCE WEEKEND 2005 CLOSES:	END

Meeting NSPKU Council of Management: (Conference Centre at 2.30pm)

NSPKU Christmas Cards

For anyone interested in purchasing the above, can you please contact **Lucy Welch** at the address below. She has this year's stock ready to go so give her a ring or drop her a line!

PO Box 26642
London
N14 4ZF

Ph: 020 8364 3010

Contacts

Dear News and Views,

Some weeks ago, my partner and I had a beautiful baby boy, (our second child). He's called Kai Mark.

Two weeks later, we were told he had PKU. We felt like our world had fallen down around us. We had never heard of PKU and everything is still new to us. We are still trying to get our heads around what has happened.

So, if there is anyone out there who would get in contact and share what they've been through and any information they have, it would be a great help.

Many thanks,
N. Wilson
53 Lang Avenue
Lundwood
Barnsley S71 5LT

NSPKU *Direct*

Help support NSPKU!

We now sell a wide range of merchandise, please use the form below remembering to state the size and colour (state a 1st and 2nd choice) where applicable. For postage and packing prices, see table below. Please fill in the form below and return to Lucy Welch at the address shown.

Lucy Welch
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Order Form NSPKU *Direct*

Product	Size	Colour	Price	Quantity	TOTAL
Adult Sweatshirts *(Red, Black, Grey, Navy, Wine)* *S. M. L. XL. XXL*		1ST CHOICE: 2ND CHOICE:	£12.00		
Adult Poloshirts *(Red, White, Navy, Jade, Grey, Wine)* *S. M. L. XL. XXL*		1ST CHOICE: 2ND CHOICE:	£9.00		
Children's Sweatshirts *(Red, Black, Grey, Navy, Wine)* *3-4, 5-6. 7-8, 9-11 years*		1ST CHOICE: 2ND CHOICE:	£8.00		
Children's Poloshirts *(Red, White, Navy, Jade, Black, Dk Green)* *3-4, 5-6. 7-8, 9-11 years*		1ST CHOICE: 2ND CHOICE:	£6.00		
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Mug 'Probably the Best Diet in the World!'			£3.50		
Christmas Cards - *Round the Christmas Tree*		10 PER PACK	£3.00 pk		
Christmas Cards - *Tree Stars*		10 PER PACK	£3.00 pk		
				+ P&P *(see below)*	£
				TOTAL REMITTANCE	£

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☐ I would like to make a donation of £: _____

Please make Cheques and Postal Orders payable to 'NSPKU'
Send completed form to:
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P+P COSTS:

Order value up to £2.00	50p
Order value £2.01-£6.00	£1
Order value £6.01-£10.00	£2
Order value £10.01-£20.00	£3
Order value over £20.01	£4

Photocopies are acceptable

All PKFoods are now available on prescription

PK FOODS

Chocolate Chip Cookies

Cinnamon Cookies

Orange Cookies

Egg Replacer

Crispbread

Pasta Spirals

Cherry Jelly Mix

Orange Jelly Mix

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Flour Mix

aminex Biscuits

aminex Cookies

aminex Rusks

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