

Welcome to  
**37th  
Annual Conference**  
2010

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**Programme Information & Abstracts**



The National Society for Phenylketonuria (United Kingdom) Ltd.

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March 2010/300

**The Rendezvous Hotel  
Skipton, Yorkshire**



The National Society for Phenylketonuria (United Kingdom) Ltd.

NSPKU, PO Box 26642, London N14 4ZF

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Please note that photographs may be taken at this event by any attendees. If you are taking photographs, please be considerate of other attendees who may appear in them. The NSPKU may use some photographs from this conference in News and Views and/or on the web site. If you have any queries regarding this matter, please contact the Conference Organizer or the Chairman.

# BOOKING FORM

## CHILDREN'S OUTINGS DURING THE NSPKU CONFERENCE 2009

For those parents wishing for their children to go on organised trips, I would ask that you please complete the details as follows and return to the conference organiser. **YOU MUST RETURN THIS FORM AS INSTRUCTED BY SATURDAY 8 AM IN ORDER FOR YOUR CHILD TO ATTEND THE TRIPS - DO NOT FORGET !**

### TRIP 1 - Saturday 6th March

**IMAX Bradford**  
(8 years and above)

Name of Child	PKU (tick)	AGE	Medical Consideration <i>other than PKU</i>

### TRIP 2 - Sunday 7th March

**Mother Shipton's Cave**  
(8 years and above)

Name of Child	PKU (tick)	AGE	Medical Consideration <i>other than PKU</i>

Children 6 and 7 years may join the trips provided they are accompanied by a parent / guardian (subject to available spaces). The parent / guardian will have to pay for their own entrance.

#### Declaration

I hereby consent for my child/children to attend the above organised trips and I/we understand that should urgent medical attention be required due to unforeseen circumstances, I/we give consent to the Party Leader to act for the child's best interests and welfare.

Signed:.....Name of Parent/Guardian.....

Date: ..... I am willing to assist with the trips. (Tick for yes)

**Please hand this form to Mike Bailey or Peter Stockton**

**NSPKU**  
**37th ANNUAL FAMILY CONFERENCE WEEKEND 2010**

**(DAY 1) – FRIDAY 5th MARCH 2010**

1430 – Welcome Registration Desk Officially Opens in  
1930 Hours the Rendezvous Hotel, Skipton:  
**Please note that no PKU / NON-PKU  
arrangements will be made  
for luncheon today**

*Location: 1st Floor, Hospitality Lounge*

1700 – Drinks Reception:  
1830 Hours **NSPKU 37th Annual Conference 2010  
will be opened by Sarah Lawson,  
Conference Organiser**

*Location: 2nd Floor, Papa Swan Bar/Hospitality Lounge*

1830 – Welcome Dinner in the hotel  
2000 Hours *Location: 3rd Floor, Malham Suite*

2000 – Evening Entertainment:  
2400 Hours **NSPKU Spring Draw 2010 plus  
DJ & Disco** *Location: 3rd Floor, Malham Suite*

**Meeting NSPKU Council of Management:**  
**9.00pm, Flasby Suite**

Important Note:- Please note that all information is correct at time of going to print.

**(DAY 2) – SATURDAY 6th MARCH 2010**

0730 – Full Breakfast will be served in the hotel:  
0900 Hours *Location: 3rd Floor, Malham Suite*

0830 – Professional Childcare Nursery  
1230 Hours **(Infants & Children under 3 years of age)**  
Professional Childcare Playgroup  
**(Children 3-7)**  
*Location: 1st Floor, Old Nursery,  
Baby Swan Restaurant*

Children's Outing: IMAX Bradford  
**{Children 8 years of age and over will be  
taken out this morning accompanied by  
adult carers} Meet at reception**

1100 – Teenagers workshop:  
1600 Hours Positive Thinking  
*Location: 4th Floor, Webber Gallery*

0920 – Opening Introduction of NSPKU 37th  
0930 Hours Annual Family Conference  
by Dave Stening (NSPKU Chair)  
*Location: 2nd Floor, Wharfedale Suite*

**MORNING SESSIONS:**

0930 – Dr. Anita MacDonald  
1015 Hours – **Weaning & research projects**  
*Location: 2nd Floor, Wharfedale Suite*

1015 – Dr. S. Huijbregts  
1100 Hours – **Executive Function in Early-treated PKU**  
*Location: 2nd Floor, Wharfedale Suite*

**(DAY 2) – SATURDAY 6th MARCH 2010** *continued*

1100 – Morning coffee/tea break & biscuits:  
1145 Hours **PKU Food manufacturers' exhibition stands  
will be open**  
*Location: 1st Floor, Papa Swan Bar/Hospitality Lounge*

1145 – Dr Kathryn Bond - **Living with PKU**  
*Location: 2nd Floor, Wharfedale Suite*

1230 – Luncheon will be served in the hotel:  
1400 Hours Raffle *Location: 3rd Floor, Malham Suite*

1400 – Firstplay Foods Demonstration  
1430 Hours  
1430 – Cookery demonstration with Eileen Green  
*Location: 2nd Floor, Wharfedale Suite*

1700– **CHILDREN'S PARTY**  
1830 Hours **11 YEARS & UNDER**  
**PARENTS TO ENSURE YOUNG CHILDREN  
ARE SUPERVISED**  
Infants/Children PKU/NON-PKU all very welcome  
*Location: 2nd Floor, Baby Swan Restaurant*

1730– **ANNUAL GENERAL MEETING:**  
1830 Hours **(EVERYONE WELCOME)**  
*Location: 2nd Floor, Wharfedale Suite*

1900 Dinner in the hotel  
2030 Hours *Location: 3rd Floor, Malham Suite*

2100 – **Pub Quiz- Live Evening Entertainment**  
2400 Hours *Location: 3rd Floor, Malham Suite*

## (DAY 3) – SUNDAY 7th MARCH 2010

0730 – Full Breakfast will be served in the hotel:  
0900 Hours *Location: 3rd Floor, Malham Suite*

0830 – Professional Childcare Nursery  
1230 Hours **(Infants & Children under 3 years of age)**  
Professional Childcare Playgroup  
**(Children 3-7)**  
*Location: 1st Floor, Old Nursery,  
Baby Swan Restaurant*

Children's Outing: IMAX Bradford  
**{Children 8 years of age and over will be  
taken out this morning accompanied by  
adult carers} Meet at reception**

0930 - **MORNING SESSIONS:**  
1015 Hours Dr Peter Galloway - **Kuvan 1 year on**  
*Location: 2nd Floor, Wharfedale Suite*

1015 - Clare Gibbings - **Travels in Africa**  
1100 Hours *Location: 2nd Floor, Wharfedale Suite*

1100– Morning coffee/tea break & biscuits:  
1145 Hours **PKU Food manufacturers' exhibition  
stands will be open**  
*Location: 1st Floor, Papa Swan Bar/ Hospitality Lounge*

1145 – 'Positive Thinking' feedback session  
1230 Hours followed by Question and Answers session  
*Location: 2nd Floor, Wharfedale Suite*

1230 – Lunch  
1400 Hours *Location: Malham Suite*

END OF NSPKU 37TH ANNUAL FAMILY CONFERENCE WEEKEND 2010

## WEANING IN PKU: THE THINGS PARENTS SHOULD KNOW!

Dr. Anita MacDonald,  
Birmingham Children's Hospital, Steelhouse Lane, Birmingham, B4 6NH

One of the things that parents of new babies with PKU worry about is weaning. Even for babies who are on a normal diet, there is no consensus about when a baby should be weaned; there is mystification about the process, as well as a multitude of commercial baby foods and equipment to choose from. Even when babies are on a normal diet, parents are taught very little about weaning, and some people may do very little cooking, so the prospect of feeding a baby on a low phenylalanine diet may be daunting. However, weaning a baby with PKU should not be too different from any other baby; it is about adapting some of the foods that are introduced, but many of the basic principles are exactly the same. The NSPKU have just produced a weaning booklet for babies with PKU which is launched at this conference. It is packed with plenty of useful weaning tips, but here are 10 additional suggestions which may be helpful.

### Getting the timing right

When you introduce any new foods, it is important that a baby is happy and relaxed so they are willing to experiment and try new things. Like everyone else, babies have to be in the right 'frame of mind' to be more experimental with their diet. Also choose a time when you are able to give plenty of time to the task and that you are unlikely to be distracted.

### Perseverance and patience

Weaning does not occur overnight. Don't be surprised if a baby does not accept a new food straight away and refuses all the pureed food you have spent hours preparing. It may take several attempts before they begin to enjoy it. So if a baby rejects a new food, try it again the following week, and again and again.

### Introduce food strategically

While most babies tend to enjoy the taste of fruit immediately (the

sweetness of fruit is similar to milk), introducing vegetables may be more difficult. It is a good idea to start with a combination of green and yellow vegetables as yellow varieties such as parsnip, butternut squash and sweet potato tend to be sweeter in flavour and can be used to soften the taste of more savoury vegetables such as broccoli. Disguising a vegetable as part of a pasta sauce or pureeing it with something they do like will help them adapt to the flavour indirectly

### **Teach by example**

Babies are more likely to try a new food if they see someone else eating it first. Before introducing new foods, it is better that adults or siblings eat some first, always making appreciative noises to show that it is being enjoyed. It may also be a good idea to give the baby a little of the new food to play with first; this is a messy but the familiarity means that they may be more likely to give it a try.

### **Make mealtime fun**

Eating regular meals at the table together with the baby (plus high chair) is a great way to make mealtimes a fun, relaxed experience rather than a chore. The baby learns more effectively by watching and imitating others, and everyone sharing at least one type of food together (maybe some free vegetable or low protein toast) will contribute to positive feeding experience. This will not only help encourage a baby's enjoyment of food but the chatter will also encourage their developmental skills.

### **Establish a routine**

Establishing a routine once weaning is started is important, as with every other aspect of dietary management in PKU. If a baby is accustomed to eating at irregular times and on demand, rather than having a routine and established daily meal schedule, it will programme them to continue this grazing pattern throughout infancy and childhood, potentially leading to stressful mealtimes.

### **Encourage baby led weaning with 'free' foods**

This is a process of self-feeding, and babies typically begin self feeding around 6 months, although some will reach for food as early as 5 months and some wait until 7 or 8 months. It should be introduced

when the baby can sit upright, is eager to participate in mealtime and may even try to grab food and put it in their mouth. It is a good way of introducing low phenylalanine foods e.g. low protein toast, pieces of soft fruit. Initial self-feeding attempts often result in very little food ingested as the baby explores textures, colours, smells and tastes, but the baby will soon start to swallow. They cope best with foods that are 'chip shaped' or have a built in handle. They can chew the bit that is sticking out of their fist and drop the rest later. As their skills improve, they will drop less food. This approach encourages independence and confidence, helps to develop hand-eye co-ordination and chewing skills, and probably reduces faddy eating and mealtime battles.

### **Keep phenylalanine exchange foods simple**

People may put so much emphasis about ensuring that the right amount of phenylalanine is eaten that it may be a precursor of long term food refusal. Sometimes babies are only offered phenylalanine exchange foods at meal time to guarantee that these are eaten. Sometimes, parents continue to spoon feed their babies because they are worried that they will leave some of their phenylalanine exchange foods. Although it is important that babies eat their daily phenylalanine allowance, in most children the number of phenylalanine exchanges allocated each day is a low number, and this is likely to remain low. Therefore, it is better to focus on offering new 'free' low phenylalanine foods at mealtime, and give phenylalanine exchange foods in the form of low volume, well tried and tested foods (e.g. cereal, yoghurt, custard), that are easy to administer.

### **What about choking?**

Many parents worry about allowing babies to feed themselves because they are concerned about choking. However, babies are at less risk of choking if can control what goes into their mouth. This is because they are not capable of intentionally moving food to the back of their mouth until after they have learnt how to chew. In turn, they do not develop the ability to chew until after they have developed the ability to reach out and grab things. It is important to always to supervise babies when eating and the size and texture of food is important.

### Babies do not read rule books

In the early stages of weaning, babies are receiving all their nutrient requirements from infant protein substitute (PKU Anamix Infant; PKU Start) and breast milk or standard infant formula. It may take time for them to eat much solid food and they will progress at their own pace. Maintaining an emphasis on play and exploration rather than on eating enables the transition to solid foods to take place as naturally as possible. What motivates a baby to make the transition to solid foods is curiosity rather than hunger. Dietitians and other professionals can give basic ground rules but it is sometimes about the parents' confidence and feeling comfortable in the process that helps the weaning process progress. The more the relaxed weaning is, the more enjoyable it will be for everyone.

## NOTES

## Executive Function in Early-Treated Phenylketonuria

Dr. Stephan Huijbregts, PhD. Leiden University, Department of Clinical Child and Adolescent Studies, Leiden, The Netherlands.  
shuijbregts@fsw.leidenuniv.nl

Executive function (EF) refers to higher-order cognitive abilities that facilitate the flexible modification of thought and behavior in response to changing cognitive or environmental demands. Examples of executive functions are planning, organization, strategy use, cognitive flexibility, inhibitory control, and working memory.

We performed a series of studies into the quality of EF in 67 children and adolescents with early and continuously treated PKU (ETPKU) and 73 controls. Results showed that children with ETPKU indeed had EF-problems, while they performed as well as controls in other areas of cognition (such as memory and motor function). Moreover, differences with controls increased when more complex EFs or combinations of EFs (e.g. inhibitory control + working memory) were called upon. The level of impairment was related to concurrent and historical phe levels. Whereas children with phe levels  $\leq 360$   $\mu\text{mol/L}$  on the day of testing performed (almost) as well as controls, those with higher levels had more difficulties. Mean phe level throughout life had the strongest relation with how well children with ETPKU did on the EF-tasks. This relation was stronger than the relation with phe on the day of testing, which emphasizes the importance of consistent dietary adherence from birth onwards. We also found more EF-problems for children with ETPKU under the age of 12 than for older children, but it should be noted that our 12-to-14 year olds generally had relatively low phe levels. Finally, our results showed that short-term dietary interventions could improve EF in ETPKU. In this presentation, these results will be illustrated, but much attention will also be given to possible new research avenues involving EF. These include follow-up studies into adolescence and early adulthood (which are usually accompanied by dietary relaxation), studies into the impact of EF-problems in daily-life of individuals with PKU, studies into brain mechanisms underlying EF-difficulties in ETPKU, and studies into the influence of new treatments for PKU on the quality of EF.

## **Living with PKU**

Dr Kathryn Bond  
Principal Clinical Psychologist in Metabolic Medicine  
Great Ormond Street Hospital  
E-mail address: bondk1@gosh.nhs.uk

PKU places many demands on families. Success with diet, supplements and blood tests can be difficult for some young people and their parents. Many young people can struggle to negotiate how to cope with feeling different from their peers. Throughout childhood and adolescence there are certain developmental stages that can be especially difficult - such as feeding young children and negotiating handing over control and independence to teenagers. This presentation will focus on the key psychological issues and strategies relevant to PKU.

**Kuvan (Sapropterin) –  
A new treatment for PKU?**

Dr Peter Galloway, Consultant Medical Biochemist  
Adult Metabolic Clinic, Glasgow Royal Infirmary

Kuvan is the first licensed medication for Phenylketonuria available in Europe for those over 4 years of age, but not for use during pregnancy.

The presentation will explain how Kuvan works and what benefits have been found. The side effects and way it is taken will be explained. Unfortunately, as a treatment only a minority benefit. How are these individuals identified and what effect could this have on their effective dietary regime?

Sapropterin is being widely used in the United States of America and in some parts of Europe. So where is Kuvan in the UK?

**Travels in Africa**

Clare Gibbings

My name is Clare-Carroll Gibbings, I am 20 years old. In August 2008 I left home for a year to do voluntary work in a South African Children's Home. As I have PKU I had slightly more to organize than most volunteers, but with support from family, friends and SHS I had a wonderful year.

Today I will be speaking about the main challenges I faced throughout the year and how I managed to cope with my diet.

I found that it was important to be honest about my condition with the charity I had applied to, and with my host in Africa. It was important to make sure everything was prepared before I left so that I could maintain my diet, but I also had to expect that things may not go as smoothly as planned.

Although I had low-protein food products delivered to Africa there were often delays at customs which meant that I did not have my supplement for a couple of months. During the year there were no major problems as I am confident with my diet but I did decide to keep a food diary for some time in order to ensure that I wasn't eating too much protein.

I was incredibly lucky to go travelling for six weeks towards the end of my year and visited Namibia, Botswana, Zimbabwe and Lesotho, as well as touring South Africa. The first three weeks of my travels were relatively simple with regards to PKU as I was on a tour with food and camping included, however the second six weeks were more difficult as I travelled independently and events were unpredictable.

After returning to Africa I continued on my diet, and I am currently reading Ancient History and Classical Archaeology at the University of Warwick.

Please complete this questionnaire so that we may make this an even better event next year.  
The Council of Management of the NSPKU

**QUESTIONNAIRE**  
**NSPKU 37th ANNUAL CONFERENCE 2010**  
The Rendezvous Hotel, Skipton

Name ..... No. in Party .....  
(optional)

Who has PKU .....

*Please tick the appropriate box for each question*

- |   | Yes                      | No                       |
|---|--------------------------|--------------------------|
| 1. Do you like the hotel staff?   | <input type="checkbox"/> | <input type="checkbox"/> |
| 2. Did you feel welcome?  | <input type="checkbox"/> | <input type="checkbox"/> |
| 3. Have you attended a conference before?   | <input type="checkbox"/> | <input type="checkbox"/> |
| 4. Did you enjoy this one?  | <input type="checkbox"/> | <input type="checkbox"/> |
| 5. Was there a good variety of topics covered?                                    | <input type="checkbox"/> | <input type="checkbox"/> |
| 6. Was the cookery demonstration informative?                                     | <input type="checkbox"/> | <input type="checkbox"/> |
| 7. What would you like to see included in future cookery demonstrations?<br>..... |                          |                          |
| 8. Were any talks too technical?<br>Which?<br>.....                               | <input type="checkbox"/> | <input type="checkbox"/> |
| 9. What topics do you wish to be covered at a future conference?<br>.....         | <input type="checkbox"/> | <input type="checkbox"/> |
| 10. Would you attend another conference?<br>.....                                 | <input type="checkbox"/> | <input type="checkbox"/> |

	<i>Very Good</i>	<i>Good</i>	<i>Ok</i>	<i>Poor</i>	<i>Very Poor</i>
11. How did you rate this conference overall?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
12. How did you rate the PKU food quality?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
13. How did you rate the PKU food choice?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
14. How did you rate the PKU food serving arrangements?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
15. How did you rate the non-PKU food?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
16. How did you rate the dining room layout & service?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
17. How did you rate the accommodation?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
18. How did you rate the creche facilities?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
19. How did you rate the creche security arrangements?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
20. How did you rate the childrens' trips?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
21. How did you rate the hotel & facilities?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
22. How did you rate the entertainment overall?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Do you have any other suggestions or comments that we may incorporate in the planning of future conferences?

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*Thank you for your time & co-operation.*

**(March 2010)**

## NOTES