

We are grateful to all the following companies who have supported us during the last year.

Firstplay Dietary Foods Ltd  
SHS International Ltd  
Vitaflo Ltd  
PK Foods  
Fate Special Foods  
YesWeCookThat.com  
Ultrapharm Ltd  
General Dietary Ltd



The National Society for Phenylketonuria (United Kingdom) Ltd.

NSPKU, PO Box 26642, London N14 4ZF

Helpline 0208 364 3010

Email: [info@nspku.org](mailto:info@nspku.org)

[www.nspku.org](http://www.nspku.org)

Charity No. 273670

© Copyright NSPKU

Company No. 1256124

February 2008/300

*Welcome to*  
**35th  
Annual Conference**  
2008

***Programme Information & Abstracts***

**The Hilton Hotel  
Northampton**

**✚ N S P K U ✚**

The National Society for Phenylketonuria (United Kingdom) Ltd.

NSPKU, PO Box 26642, London N14 4ZF





# Contents

|  |              |
|--|--------------|
| <b>Childrens Outings Booking form .....</b>  | <b>3</b>     |
| <b>Hotel Floor Plan .....</b>  | <b>5</b>     |
| <b>Conference Programme .....</b>  | <b>7-10</b>  |
| <b>Being you with PKU .....</b>  | <b>11</b>    |
| Danuta Orłowska Clinical Psychologist London .   |              |
| <b>Obtaining Special Low Protein Foods:<br/>A Door to Door Service .....</b>                                 | <b>13-15</b> |
| Anita MacDonald, Anne Daly, Vanessa Hopkins  |              |
| <b>Adults with previously untreated phenylketonuria (PKU)<br/>– a randomised control trial .....</b>         | <b>17-18</b> |
| Lesley Robertson   |              |
| <b>A Hundred Ways with Vegetables and<br/>Making a Recipe Work for You. ....</b>                             | <b>19</b>    |
| Barbara Cochrane Senior Dietitian Yorkhill Children’s Hospital Glasgow<br>and Eleanor Weetch NSPKU Dietitian |              |
| <b>Talking about PKU across the life-span .....</b>  | <b>21</b>    |
| Dr Peter Burgard. Centre for Paediatric and Adolescent Medicine, University of<br>Heidelberg – Germany       |              |
| <b>“It doesn’t have to be like this...” .....</b>  | <b>23</b>    |
| Joe O’Malley   |              |
| <b>Questionnaire .....</b>   | <b>25-27</b> |

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.

The views expressed in these papers are those of the authors and not necessarily those of the NSPKU.



# BOOKING FORM

## CHILDREN'S OUTINGS DURING THE NSPKU CONFERENCE 2008

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 1st March

**Kids Play**  
(8 years and above)

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

### TRIP 2 - Sunday 2nd March

**Westlodge Rural Centre**  
(8 years and above)

| Name of Child | PKU<br>(tick) | AGE | Medical Consideration<br><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

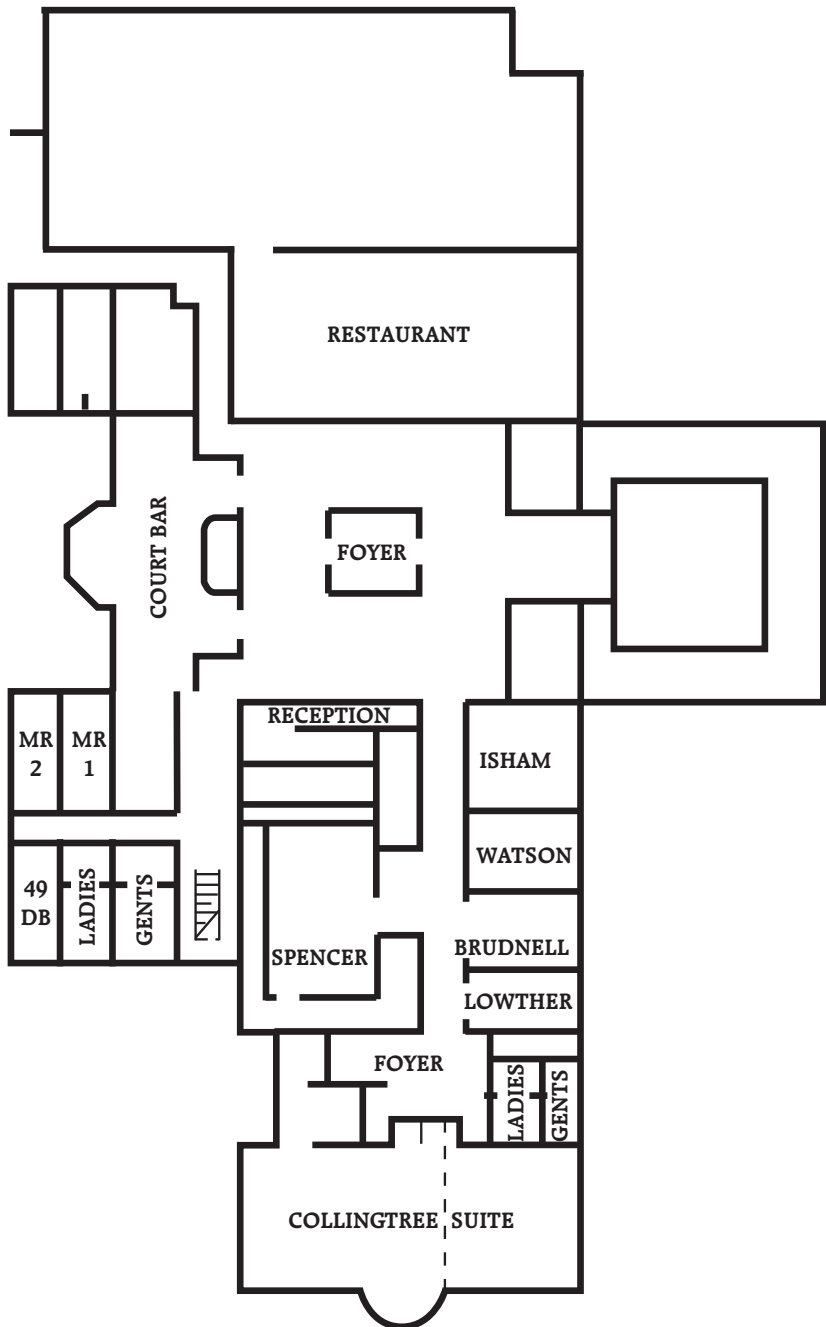
I hereby consent for my child/children to attend the above organised trips and I/we understand that should urgent medical attention be required due to unforeseen circumstances, I/we give consent to the Party Leader to act for the child's best interests and welfare.

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

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

**Please hand this form to The Conference Organiser**







**NSPKU**  
**35th ANNUAL FAMILY CONFERENCE WEEKEND 2008**

**(DAY 1) – FRIDAY 29th FEBRUARY 2008**

1430 hrs      NSPKU 35th annual family conference  
weekend registration desk officially opens in  
the hotel

*Please note that no PKU/NON-PKU  
arrangements will be made for luncheon today*

Remainder of afternoon at guests' own leisure

1700 hrs      Welcome drinks reception, Court Bar

1815 hrs      Welcome dinner, restaurant

1945 hrs      **Evening entertainment programme:**

NSPKU 2008 Spring Draw & DJ & Karaoke  
Collingtree Suite

**(DAY 2) – SATURDAY 1ST MARCH 2008**

- 0730-0845hrs** Full English breakfast will be served.  
Restaurant
- 0830 hrs Professional nursery/playgroup  
Spencer Room  
(infants & children under 8 years)
- Children's outing: meet reception  
*(children 8 years and over will be taken out  
this morning with adult carers)*
- 0900 hrs NSPKU 35th Annual Family Conference  
weekend 2008: Collingtree Suite  
opening introduction,  
David Stening – NSPKU Chair
- 0915 hrs Being You with PKU –  
Dr. Danuta Orłowska, Clinical Psychologist
- 0930 hrs Teenagers Outing (13-19 years old)  
GP Go Karting.  
Minimum height 1.45m (4ft 9in,
- 1000 hrs Research Update – Dr. Anita MacDonald
- 1045 hrs** Morning coffee/tea break & biscuits  
(PKU food manufacturers exhibition open)
- 1115 hrs Untreated Trial –  
Lesley Robertson, Janet & Carolyne
- 1215 hrs New Products –  
Eleanor Weetch, Barbara Cochrane

**(DAY 2) – SATURDAY 1ST MARCH 2008** *continued*

- 1300 hrs Luncheon will be served, Restaurant Raffle
- 1400 hrs Cookery demonstration, Collingtree Suite with Eileen Green Fate Foods
- 1700-1830 hrs Children's (PKU and non-PKU) party, Isham (11 years & under– Parents to ensure young children are supervised)
- 1730 hrs 2008 NSPKU Annual General Meeting – Collingtree Suite All Welcome
- 1830 hrs PKU Teenagers and Young Adults Gathering Brundenell Room – your chance to say what you want
- 1930 hrs **Final dinner**, Restaurant
- 2030hrs **Pub Quiz** Collingtree Suite
- 2100 hrs **Evening live entertainment** Collingtree Suite

**(DAY 3) – SUNDAY 2nd MARCH 2008**

**0730-0845hrs Full English breakfast, Restaurant**

0830 hrs Professional nursery/playgroup,  
Spencer Room  
*(infants & children under 8 years)*

Children's outing: Meet Reception  
*(children 8 years and over will be taken out  
this morning with adult carers)*

0915 hrs PKU across the Life-Span, Collingtree Suite –  
Dr Peter Burgard

1000 hr It doesn't have to be like this –  
Fiona & Joe O'Malley

**1045 hrs Morning coffee/tea break & biscuits**  
(PKU food manufacturers exhibition open)

1115 hrs 'So you think you know your exchanges'

1200 hrs Question Time

**1230 hrs Luncheon, Restaurant**

**END OF NSPKU 35TH ANNUAL FAMILY CONFERENCE  
WEEKEND 2008**

PLEASE NOTE THAT THIS IS A PROVISIONAL PROGRAMME  
OF EVENTS AND IS SUBJECT TO CHANGE

# **Being you with PKU**

Danuta Orłowska Clinical Psychologist London

This talk is in three parts.

The first part considers our mental well-being: what is it and how can we look after it? There will also be a brief discussion of ideas and tips.

The second part introduces ideas drawn from cognitive therapy. It shows how these can help tackle the unhelpful ways of thinking which are often linked with distress.

The final part shows how cognitive therapy skills can be applied to living with PKU (whether on or off diet). The skills can also be used in helping us deal with challenges and goals we set ourselves whether health-related or not.

# NOTES

# **OBTAINING SPECIAL LOW PROTEIN FOODS: A DOOR TO DOOR SERVICE**

Anita MacDonald, Anne Daly, Vanessa Hopkins

Birmingham Childrens Hospital, Steelhouse Lane, Birmingham, B4 6NH

## **Background**

Over the last 4 years, patients with PKU have been able to obtain their phenylalanine-free protein substitutes and low protein milks (e.g. Sno Pro, Duocal) via one of two home delivery companies. This type of service has been greatly appreciated by the families who have chosen to use it. It has guaranteed a consistent supply of these essential products providing there has been regular communication with the home delivery team. The latest challenge was to examine if this type of service could be extended to other low protein products.

## **Why have low protein products on home delivery?**

In the UK, there are approximately 120 special low protein foods, which are generally only available through the NHS system. This means they have to be prescribed by the GP, and then obtained via a chemist. This system has remained unchanged for over 30 years, even though the range and choice of special foods has greatly expanded. This system works well for some families; but not others. Some families keep to the same low protein products (e.g. bread, pasta, and flour); whereas others want more variety and like to try all the low protein products available, but many GP practices dislike altering prescriptions from month to month. Some families feel belittled and guilty requesting low protein foods on prescription. The system is not very dignified and they may feel they are begging for food. Sometimes low protein foods arrive at the chemist in the wrong quantity e.g. twelve boxes instead of one. Any new system that aims to avoid some of these difficulties is worth evaluation.

## **What is the home delivery system for low protein foods all about?**

This is a service that is offered by SHS Nutricia for their low protein foods (and protein substitutes) only. SHS Nutricia produces about 35%

of all low protein special foods. They also make special low protein milks and protein substitutes. This new service has been piloted in 3 PKU centres; Belfast, Birmingham and Glasgow.

### **How does it work?**

In the West Midlands, the home delivery company sends patients/parents a monthly picture order sheet for all of the low protein products and special low protein milks made by SHS Nutricia. This form takes seconds to complete, patients/parents return this form to the home delivery company who then request a prescription from the GP. The low protein special foods are delivered to the family home within 28 days of the original order sheet being completed.

### **What are the advantages?**

There are many benefits to the system. Families have a guaranteed supply of product providing they follow the simple procedure; they are likely to be able to access a variety of products; the right special foods in the right amounts are likely to be delivered; it is convenient; less contact is needed with the GP practice; and generally the families have been delighted with the service. It has been particularly beneficial for new families, working parents or patients, late treated patients in care homes and pregnant women with PKU.

### **What are the disadvantages?**

Parents/families clearly have to plan ahead, and think about what they are likely to need four weeks in advance. Some families do not return their order forms on time but they only have one chance to return their order forms each month; and if they miss the deadline that is it for another month. A few GP practices are inflexible about prescribing different products; whereas some distrust the new system. Some families would prefer a few standard products e.g. low protein pasta or bread delivering each month rather than having to choose products.

### **What about using products from other companies?**

The patients/parents order these in the same way as before from their GP. This does involve the GP issuing more than one prescription but many people tend to order other products in larger quantities so they minimise prescription requests. Once the protein substitute and low

milks are delivered monthly by the home delivery service, it is easier to order other low protein foods once every two to three months.

### **Is this service Eutopia?**

Not yet; but it is a good start. The perfect service would be inclusive of all the low protein products; allow patients/parents to obtain low protein products without the consent of their GP, take care of the prescriptions of vulnerable families and patients, minimise any wastage but still ensure that everyone has a generous supply of everything they need. Therefore, we have not yet reached Eutopia, but we are on the right road!

# NOTES

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

Lesley Robertson

## **Introduction**

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

## **The Study**

The 36 participants were each studied over a period of 60 weeks. Initially there was an 8 week baseline followed by two 24 week trial diets (low and normal phenylalanine diet) with a four week washout period between them.

All were monitored at regular intervals throughout the 60 weeks, this included weekly blood phenylalanine and a range of behaviour monitoring.

## **Results**

Of the 36 participants, only 17 completed the full 60 weeks. 13 withdrew in the baseline or the first trial diet due to the dislike of the amino acid product or blood test refusal. The remaining 6 withdrew for different reasons at various stages.

## **Blood Results**

Blood phenylalanine was monitored weekly throughout the trial with good control during the active low phenylalanine diet. As expected during the other phases of the trial blood phenylalanine was high.

## **Behaviour Results**

Behaviour monitoring was carried out 5 times during the study – at the very beginning and at the end of each phase. This consisted of standardised questionnaires and 3 short video's per visit. The analysis of

these showed no significant differences during the active diet. A daily behaviour diary was completed by care staff. This recorded 3 positive, 3 negative behaviours and carer comments. No significant differences were noted with positive and negative behaviours. However carer comments showed a significant benefit in the active phase.

### **Conclusions**

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

### **Acknowledgements**

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

# **A Hundred Ways with Vegetables and Making a Recipe Work for You.**

Barbara Cochrane Senior Dietitian Yorkhill Children's Hospital Glasgow  
and Eleanor Weetch NSPKU Dietitian

You will be working with dietitians and other delegates to open your mind to the versatile ways you can present all kinds of vegetables – the common and the not so common. This is going to be an interactive session where we help one another.

Recipe adaptation will also be covered so you needn't discard a recipe as unsuitable – you can learn how to make it work for you.

Useful products in the supermarket will also be incorporated into the session.

# NOTES

# Talking about PKU across the life-span

Dr Peter Burgard

Centre for Paediatric and Adolescent Medicine  
University of Heidelberg – Germany

Phenylketonuria (PKU) is a rare condition, for lay people as well as for paediatric specialists. Talking about the diagnosis, treatment and management is essential to establish successful and trustful relationships between parents, patients and professionals. Communication about PKU includes concepts and terms not natural in everyday language or in medical terminology. In addition talking about PKU to children and adolescents has to be tailored to their developmental age. Talking about PKU deals with bringing thoughts to action, ie to compliance with recommendations for treatment. Last but not least talking about PKU must take into consideration emotions, ie anxiety, helplessness, guilt, shame, and embarrassment but also about surprise, luck, hope, joy and coolness. Talking about PKU across the life span also has to deal with the scientific evidence for treatment and management during infancy, childhood, adolescence and adulthood.

Based on my experience for nearly 20 years of counselling professionals (general paediatricians, paediatric metabolic specialists, dietitians), families and patients with PKU I will present examples across the life-span how communication can be improved in order to support coping with the condition and to increase the medical power of information.

# NOTES

## **“It doesn’t have to be like this...”**

Joe O’Malley

Fiona didn’t realise at the time the significance of that sentence.

As a child Fiona often received glowing school reports citing her willingness to learn and be helpful together with a sunny outlook. She was a perfectly behaved pupil; the only thing that set her apart from other children was what she would have for lunch.

Once Fiona started working and was in full control of her own eating habits the years of being good allowed her a certain ‘discretion,’ the occasional extra exchange, birthday treat and Christmas allowance. Gradually the treats and extra exchanges became blurred and merged with the routine of work. When her Dietician asked why her phenylalanine levels were so high Fiona wasn’t sure, she had been taking the supplements and her diet wasn’t that far off track...

The denial and self assurance came to a head one Saturday morning in October 2005 when Fiona was found innocently enjoying a full English breakfast.

Fiona has phenylketonuria. At that time she had phenylalanine levels in excess of 2,000  $\mu\text{mol/l}$ , weighed in excess of 200lbs and had a BMI of 36.6. These were the measurable elements of Fiona’s decline. What medicine couldn’t measure were the associated headaches, irritability, mood swings and depression that compounded her situation.

Returning from university it was her brother who noticed just how much Fiona had changed. After a lot of listening, research and an idea it was he who said “It doesn’t have to be like this...”

This is Fiona’s story. This is how far she has come.

# NOTES

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 35th ANNUAL CONFERENCE 2008

### The Hilton Hotel

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"/> |

**Very Good**

**Good**

**Ok**

**Poor**

**Very Poor**

11. How did you rate this conference overall?

12. How did you rate the PKU food quality?

13. How did you rate the PKU food choice?

14. How did you rate the PKU food serving arrangements?

15. How did you rate the non-PKU food?

16. How did you rate the dining room layout & service?

17. How did you rate the accommodation?

18. How did you rate the creche facilities?

19. How did you rate the creche security arrangements?

20. How did you rate the childrens' trips?

21. How did you rate the hotel & facilities?

22. How did you rate the entertainment overall?

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

.....

.....

.....

.....

.....

.....

.....

.....

.....

.....

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

**(March 2008)**

# NOTES