

The British Porphyria Association newsletter



Registered Charity No.1089609

ISSUE 16 • MAY 2008



Calling all skin Porphyria sufferers...

Let's face it, last year's summer may not have been so popular with the rest of the population but it was great for us!! In case it's a sunny one this year, we need to start thinking about buying the sun cream and covering up.

Normal sunscreens don't offer enough protection especially in the case of EPP sufferers, who need a reflectant sunscreen as they cover both UVA, UVB and visible light. Examples of these types of products available at chemists are:

- Ambre Solaire® lotion SPF 60
- Delph® lotion SPF25
- Sensense® Ultra SPF 60
- Delph® lotion SPF 30
- E45 Sun® lotion SPF25
- Ultrablock® cream SPF30
- E45 Sun® lotion SPF50
- Vichy factor 60A

If you haven't already used Dundee Cream it's worth a try. Ask your GP to prescribe it to for you. Your GP/chemist will need to contact Dundee Pharmaceuticals, Ninewells Hospital, Dundee. DD1 9SY, telephone: 01382 632052. Dundee Cream comes in three colours : beige, white and coffee.

Remember the Australian saying! 'slip, slap, slop' is relevant to everyone but definitely for us. It's best to wear long sleeves, a hat, gloves and glasses. (And good news - apparently the pasty look is in this year!)

For further useful information check out British Association of Dermatologists website www.bad.org.uk and type in Porphyria on the top left search bar.

UV protective clothing which may be useful can be found on websites www.suninsight.com and www.equatorsun.com

Disclaimer – BPA do not endorse any products. These are only suggestions and, in the case of EPP, UV protective clothing will not offer full protection.

Introducing the UK Acute Intermittent Porphyria Study (UK AIP)

Do you have AIP?

Does someone in your family have AIP?

Researchers from the National Porphyria Service in Cardiff and from Cheshire & Merseyside Clinical Genetics Service will soon be looking for patients and family members willing to take part in a national research study into AIP.

Our aim is to try to understand why AIP affects some people more severely than others. It is important to understand this in order to develop improvements in treatment and advice given to individuals and families at risk of AIP. A national study of this kind has not been undertaken before in the UK and we hope that as many families as possible will be able to take part.

Taking part in the study is likely to involve a one-off telephone call from a researcher; some people will also be asked to meet with the researcher at their nearest participating hospital centre.

We plan to start recruiting for this study in Autumn 2008. If you or your relative has AIP and you would like to find out more about the study in the meantime, please contact Dr Helen Murphy, c/o Dept of Clinical Genetics, Alder Hey Hospital, Eaton Road, Liverpool, L12 2AP, Tel: 0151 802 5031, Fax: 0151 802 5096, Email: lwt.AIPstudy@nhs.net.

We are grateful to the British Porphyria Association for supporting this study. More information will be also available at the BPA day in Leeds on 1st June 2008 and at the BPA AGM in September in Liverpool.

www.porphyria.org.uk



BPA AGM and Conference

This year we had our own AGM at Cardiff. Our committee has remained the same.

Thanks to the fund-raising of friends of Helen Gibbs, we have more than £6,000 available to prime research on the acute porphyrias.

We have since decided to use half of the Helen Gibbs fund - plus some from our general fund - to support the research work into the effects of AIP by Dr Helen Murphy, which is described on the front page.



We asked Dr Mike Badminton to be a Patron of the BPA, and he accepted.

Our first speaker after the AGM was Dr Ruwani Katugampola, who is doing a study on CEP partly funded by the BPA. CEP is a "skin" porphyria caused by a recessive gene (2 copies have to be inherited, one from each parent). It is rare, so the study took a long time to set up, getting permission from many different hospitals and doctors – lots of forms to fill in.

The aims of the study were to assess quality of life, to look at the gene (DNA) to see if the position of the fault could predict the severity, and to highlight issues for affected families (such as the difficulty of accessing disability benefits). One investigator (Ru herself) did the examinations and asked the questions at the local hospitals, but lab investigations were done at Cardiff.

CEP can cause disfigurement in severe cases. Affected babies can have red urine, and easily blister on light-exposed areas, so it is sometimes seen early on, though some only develop problems in their 30s or 40s. Life can be difficult for those badly affected, having to cover up or use special creams, and some needing blood transfusions which can cause iron overload. It disrupts normal family life, making things difficult for both the parents and other children.

It also limits the choice of job in adult life. A bone marrow transplant can cure CEP, but this is not without risk. Ru has completed the study, but there is still a lot of work to do, particularly the final report.

Dr Mike Badminton then spoke about developments. He mentioned a previous study on EPP, which has proved useful in genetic testing – if the fault in a parent's gene is known, testing the children is very quick. Ru is just completing the study on CEP and there is one on AIP in the planning stage (Cardiff and Liverpool).

Commercially, there has been a study of Enzyme Replacement Therapy for AIP, but unfortunately there was no benefit found. A study is about to start on the use of a hormone in EPP to stimulate the production of brown pigment in the skin. Tanning, using carefully controlled exposure, is known to help those with EPP, but it needs regular access to a hospital with the right equipment, so it is hoped the hormone treatment will also help.

Mike also explained about EPNET and the work needed to compile the porphyria-europe web site. This has information for both patients and doctors on acute porphyrias, and information on non-acute porphyrias is gradually being added. He also encouraged "acute" members to report on drugs to Susan George at WMIC (see separate article).

We then had a question and answer session with Mike and Ru. Summary of points:

Fatigue:

Mike commented that this is often reported by those with an acute porphyria. It is difficult to assess, but after an attack there may be problems caused by diet or lack of food, and by the drugs which have been needed.

Defining an acute attack:

This is very difficult. Even the experts can't do it!

Safe drugs for acute porphyria:

One member had been given an "unsafe" drug by her doctor, even though she had queried it. It caused an attack. Mike recommended that she always check drugs herself, rather than relying on a doctor to check.

How long does haem arginate last after treatment?:

After 4 doses, the concentration in the blood reaches a steady state briefly. It then falls off fairly quickly. It may persist longer in the liver, but no-one knows.

Alcohol and diet:

Alcohol is damaging to the liver, so it's best avoided by everyone, or at least taken in moderation. "Superfoods" are likely to be good for you.

EPP and anaemia: Most with EPP seem to be about 11/2 g/dl low on iron. Their bodies have probably adjusted to the lower level, so they are not strictly anaemic. If iron is given, tests should be done to check if the blood level rises. If not, then it isn't needed. Some report that iron makes their skin more sensitive, but others have no problems.

Subs for BPA

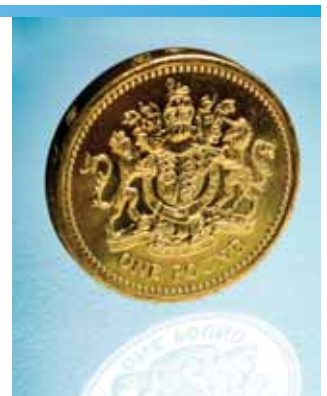
A part of our income comes from your subscriptions.

This is used for running the BPA (e.g. producing this newsletter), helping our members, and supporting research. We ask for £10 pounds per family as a minimum, if you can afford it. **If you can give more, please do.**

Can we thank all of you that have sent us your subs this year. If you haven't, please do so now! Sending out chasing-up letters diverts funds from better uses of them!

Several of our members pay by standing order.

Have you thought of setting up a standing order to pay us say £2 a month?



Internet Fables.

According to a site on the internet, there are a lot of foods that porphyrics shouldn't be eating. One of our correspondents got very hot under the collar about broccoli, claiming it had caused her acute attacks.

We can only assume that she is actually allergic to broccoli, since all the "acutes" on the committee (and "acute" members of their families) eat broccoli regularly. One, who has had an attack, says he's very fond of broccoli and eats it frequently with no ill effects!

There is no evidence that food or drink trigger porphyria symptoms, except for alcohol, and the specific triggers for PCT (which PCT patients should already know about).

As with any illness or discomfort, don't assume (or let your doctor assume) that anything new is always due to the porphyria. Appendicitis isn't! Some people with a porphyria also have food sensitivities or allergies (just like those without a porphyria) and these can get worse as you get older. If you're not sure of the cause, cut out one thing at a time from your diet – for example, if it might be cheese, cut out cheese for a week, then eat some again and see if the symptoms return. If that wasn't the cause, try cutting out something else.



If it proves difficult to track down, keeping a food diary may help. Note what you eat each day and when you feel bad. It may also be worth noting where you go - a friend recently had a very nasty skin reaction to some toilet cleaner which had just been used in a public toilet, and my mother got hay-fever-like symptoms from some scents and from fresh paint. Noting where you've been may be enough to give you a clue to the cause.

The internet is a very useful source of information, but do use it with caution. Unscrupulous people will try to sell you "cures" which do nothing. Others will just misunderstand evidence and perpetuate myths. The BPA was founded to give you reliable information so, if in doubt, do write to us or give us a call.

Kirstine Chamberlayne

FUNDRAISING

As a small charity we have to rely on the generosity of members and their families to give their time and effort to help us to raise the funds needed to: run the charity, e.g. telephone, website; support research projects, which ultimately help us all; and to support members and their families emotionally, and financially if possible.

We have included a sheet of FUNDRAISING ideas with this newsletter. Please take time to look through the list and find something that you could do to help. You may be able to encourage friends, work colleagues and family members to participate too.

We are asking everyone to participate in our Porphyria Awareness Day on or around Sunday June 1st by holding a coffee and cakes morning, a car washing afternoon, or a cheese and wine evening. Some of you may even be able to do ALL three! If you work in an office, why not play Fifty to One. Take one fifty pound note and seal into an envelope. Mark a large sheet of paper (A3 is best) into 100 boxes, numbered from 00 up to 99. Ask work colleagues to 'buy' one square for £1. When all of the squares have been bought, open the envelope. The person who bought the square that contains the last two numbers on the note wins the note, and the remaining £50 goes to the BPA.

We can help you by providing some helpful advice to make your fundraising easy and fun. Phone our helpline on 01474 369231.

Fundraising for The British Porphyria Association...

Choice, savings and value for you, raising money for us at the same time!

Visit www.buy.at/BPA
for more great offers from over 100 top retailers.



You can raise over £300 for The British Porphyria Association just by shopping through www.buy.at/BPA.

Retailer	Commission	
An ISA from Legal & General	£50	
Sky HD package	£60	
A contract phone from Dial-a-phone	£60	
Home insurance from the CO-OP	£70	
AA UK breakdown	£30	
Free 21 day DVD rental trial from LOVEFILM	£7.50	
Egg card	£20	
National Trust membership	£20	
£150 on clothes from All Saints	£15	
Total	£332.50	

www.porphyrria.org.uk

HELPLINE

We are considering moving our help-line number to a free-phone number, but for the time being it remains:

T: 01474 369 231

Would members please note that our helpline is only manned at specific times, due to work and family commitments.

When it is not manned, an answer machine will be in operation.

Please leave name, phone number, day and time of message and someone will call you back as soon as possible.

Web: www.porphyria.org.uk

E-mail: helpline@porphyria.org.uk

**Address: BPA, 136 Devonshire Rd
Durham City, DH1 2BL.**

The BPA Newsletter is published by the BPA twice yearly.

Chairman: John Chamberlayne

Treasurer: Alan Molyneux

Secretary: Sarah Pepperdine

Patrons: Prof. Timothy Cox, Prof. George Elder
Dr. Mike Badminton

View points and opinions contained in this newsletter are reproduced in good faith and do not necessarily reflect the judgement of the British Porphyria Association or its patrons.

Hospital Open Day

We are having another open day this year, organised by Dr Julian Barth and Keith Allen at Leeds. The open day will be at Britannia House, Britannia Road, Morley, Leeds, LS27 0DQ, on **Sunday 1st June 2008**.

The provisional programme:

10.30 am	Coffee
11.00 am	Welcome by Keith Allen, Consultant Clinical Scientist.
11.15 am	News/update on the BPA, by John Chamberlayne, BPA chairman
11.30 am	The AIP Study, by Dr Helen Murphy, Liverpool
12.15 pm	Photoprotection and the skin, by Dr Alison Layton
12.45 pm	Role of the Laboratory, by Dr Elizabeth Fox, Biochemist, Leeds
1.15 pm	Lunch followed by Laboratory tour (in groups)
3.15 pm	Question & Answer Session
4.00 pm	End

Lunch/Coffee will be provided by courtesy of Orphan Europe UK Ltd.

There is ample parking.

Previous open days have proved to be very popular, so do come along.

If you are hoping to come, please let the BPA know, and we will send full details of how to get there.

Autumn Half-day Conference and AGM 2008

We are having a half-day conference in **Liverpool, on 27th September**.

It will start at 2.00pm and be at Liverpool Women's Hospital, Crown St, Liverpool, L8 7SS.

After a short AGM, we will have a talk by Dr Felicity Stewart of Salford Royal (Hope) Hospital, and one by Dr Helen Murphy, who is doing research into AIP at Liverpool.

This will be followed by a discussion on porphyria clinics, and then a question and answer session.

We will then provide a buffet tea, when you can talk to other porphyria patients, to the committee, and to the medical experts.



British Porphyria Association Registered Charity No. 1089609

MEMBERSHIP / DONATION FORM & STANDING ORDER

TITLE: _____ NAME: _____

ADDRESS: _____

POSTCODE: _____

EMAIL: _____

TELEPHONE: _____

TYPE OF PORPHYRIA: _____

DATE: _____

WAYS TO PAY

- I would like to pay my annual membership fee of £10
- I would like to make a donation of:
 £10 £20 £30 £40 £50 other £ _____
- I would like to pay by Standing Order (please fill in the form opposite)
- I enclose a cheque made payable to the 'British Porphyria Association' for £ _____
- I would like a receipt

By making a donation to the British Porphyria Association you will know that you are making a vital contribution to our work. Simply making a small donation will help us raise awareness, expand our help line or support our research.

DO YOU PAY UK TAX?

If you do, you can add 28% to the value of your gift to the British Porphyria Association at no extra cost to yourself. Simply sign and date the Gift Aid declaration and for every £1 you give we can ask the Inland revenue to give us an extra 28p.

Standing Order BRITISH PORPHYRIA ASSOCIATION

Please send this form to: The Treasurer, British Porphyria Association, 599 Bolton Road, Aspull, Wigan, Lancs, WN2 1PZ.

A monthly standing order or any donation you can give will make a difference.

BANK/BUILDING SOCIETY NAME: _____

BRANCH ADDRESS: _____

POSTCODE: _____

Please pay the British Porphyria Association the sum of £ _____ each month/quarter/year (delete as appropriate) from my account until further notice.

ACCOUNT NAME(S): _____

SORT CODE: _____ ACCOUNT NO: _____

STARTING ON* (DATE): _____

*This date must be more than one month after today's date

SIGNED: _____ TODAY'S DATE: _____

This cancels all existing Standing Orders to the British Porphyria Association (please tick) yes no not applicable

BPA use only: Please pay to British Porphyria Association bank account:

20-43-63 _____

GIFT AID DECLARATION

I am a UK Tax payer and would like the British Porphyria Association (Registered Charity No. 1089609) to treat all donations I have made since 6th April 2000, and any future donations, as Gift Aid donations until I notify you otherwise. (We can reclaim the tax on your donations at no cost to yourself.)

DATE: _____ SIGNATURE: _____