



## 3rd AGM of the BPA

### 24th October 2004

The AGM of the British Porphyria Association was held on Sunday 24th October 2004 at Shirley Methodist Church, Birmingham.

There is a considerable change this year to the committee of the BPA, so we voted in the new committee.

John Chamberlayne (chair@porphyria.org.uk) was voted as the new chairman; and Leigh Drake (secretary@porphyria.org.uk) as the new correspondence secretary.

We also elected the following officers, unopposed: Ann Newton (treasurer@porphyria.org.uk) as Treasurer, Kirstine Chamberlayne as minutes secretary, Sarah Clark as PR & marketing, and Karen Harris to remain manning the help-line.

It was announced that subscriptions would be collected from all members for the year at the beginning of each year. All members will be receiving a letter in January.

There was discussion regarding getting one's GP to refer porphyria patients on to specialists.

We were pleased to welcome Dr Alex Holme, who is currently working on research into liver disease in EPP patients. He gave an interesting talk about his research. Following that we presented him with a cheque for £2000 towards his research.

Following the formal AGM there were informal discussions and a chance to talk to other members.



# NON-PORPHYRIC SKIN DISEASE IN PORPHYRIA PATIENTS

**ACNE:** Topical treatments for mild to moderate acne are generally safe in patients with porphyria, though there is scant evidence in the literature. Unfortunately, none of the commonly-used antibiotics are safe. Similarly, Isotretinoin, the most effective treatment for acne, is unsafe in the acute porphyrias and should be avoided.

**ECZEMA:** Eczema is often controlled with a combination of moisturizers and steroid creams or ointments. These treatments are generally safe. Moisturizers keep the skin in good condition, which may help to avoid minor injuries of the skin which may lead to scarring. Anti-histamine tablets are sometimes given to help reduce itching. Chlorpheniramine (Piriton) and Promethazine (Phenergan) are considered safe. Most other anti-histamines should be avoided. Considering systemic treatments for eczema, Prednisolone and Azathioprine are both safe in all forms of porphyria. Ciclosporin, another agent occasionally used to treat eczema, is not safe. For ultraviolet treatment of eczema, see below under psoriasis.

**PSORIASIS:** Treatment with moisturizers and steroid creams/ointments is safe. Topical treatments such as Calcipotriol, Dithranol and coal tar are not known to be harmful. Systemic treatments for psoriasis include Methotrexate which must be used



with caution. Acitretin and Ciclosporin are both unsafe. Ultraviolet light treatments are often used together with psoralens tablets for both psoriasis and eczema, however it should be avoided by patients with photosensitive porphyrias. Psoralens tablets which are known to be unsafe should be avoided by those who have any of the acute porphyrias.

**SKIN SURGERY:** Sometimes the skin needs to be sampled either to help make the diagnosis or to remove benign or malignant tumours. This is usually done under local anaesthetic. Most local anaesthetic agents are safe in porphyria, but Bupivacaine (Marcaine) is preferred.

**WARTS:** Preparations such as salicylic acid (cream or paint) are safe but care should be taken since the skin in porphyria is fragile. This is also the case for cryosurgery.

## PROTECTIVE SUNSCREENS & PROTECTIVE CLOTHING

We are pleased to advise patients with EPP, PCT & VP that we have negotiated a 10% discount off retail prices (below) with TYPHARM LTD, suppliers of suncreening products by Senses.

### Products available are:

Daily Face (SPF 60)/75g/£10.58  
Low Irritant (SPF 20)/125ml/£8.99  
Sport Gel (SPF 30)/125ml/£8.99  
Toddler Milk (SPF 50)/50ml roll on/£5.49  
Toddler Milk (SPF 50)/125ml/£8.99  
Ultra (SPF 60)/50ml roll on/£5.49  
Ultra (SPF 60)/125ml/£8.99

Members should ring Typharm on 01603 735200 and quote The British Porphyria Association. The price includes postage & packing.

### SunSibility UV Protective Clothing

Clothing offering SPF 50 is also now available from 'SUNSIBILITY' UV PROTECTIVE CLOTHING, and they too will give a 10% discount to members quoting The British Porphyria Association.

SunSibility Clothing is only available by mail order and is delivered straight to your door! They produce flattering designs in sizes 8-22 for women, small to XX large for men and ages 0-16 for childrenswear, in a multitude of colours and innovative designs as well as accessories complementing the collection.

Those members known to have PCT, EPP or VP should have received a coloured leaflet with further details. Alternatively, they can be contacted by phone, post, e-mail or via their website:

3 Chadacre Road, Stoneleigh,  
Epsom, Surrey, KT17 2HD  
www.sunsibility.co.uk  
sunsibility@hotmail.com  
Telephone 0208 224 0834.

# The BPA web site

[www.porphyria.org.uk](http://www.porphyria.org.uk)

- An introductory page, about the association;
- A news page - currently giving information about our AGM, and our newly elected officers.
- A question and answer page on acute porphyrias
- A form for donating to the BPA !
- Contact details for the BPA, and its officers.
- Contact details of Porphyria experts in the UK, and world-wide
- Links to other sites relevant to those with Porphyria. We will be adding to this.

We will be adding to it steadily over the coming months. As well as keeping the news pages up-to-date, current plans include information on member's experiences, and suggestions for dealing with common problems with Porphyria.

Suggestions for additions or improvements for the web site are very welcome. Phone the help line, or better, email [webmaster@porphyria.org.uk](mailto:webmaster@porphyria.org.uk).

## MEMBERSHIP FEES 2005

### Q. What do membership fees cover?

#### A. Membership fees cover the following:

- a) Newsletter (though some have been sponsored)
- b) Administration costs, including the helpline, which costs approx £250 pa at present. Stationery, printer ink, postage, envelopes.
- c) Travel costs of committee/trustees on BPA business or to attend meetings on behalf of the BPA. (Members travel to BPA committee meetings approx 3 times a year, which are currently held in different locations across the UK so that the same members do not always have to do long journeys. We pay mileage at 15p/mile.)
- d) Overnight accommodation of committee/trustees if undertaking business or to attend meetings on behalf of the BPA which requires the member to stay overnight.
- e) Any money above these needs will be put towards the money we are raising for help to members and support of research into porphyria.

### Q. How many members are there?

A. Currently 255 members are registered with the BPA

### Q. How much income is actually generated from membership fees?

A. For the year Jan 2004- Sep 2004 ONLY £130 of membership fees have been paid by cheque, plus approx. £60 by standing order. (Not even enough to cover the telephone bill this year)

### Q. How much could be generated by membership fees?

A. Membership fees are set at £10 per person and based on our database this should produce £2550 per annum.

### Q. When are membership fees due, and will I get a reminder?

A. With effect from January 2005, all membership fees will be due in advance and a reminder will be sent out in January

### 5. How do I set up a standing order, and can this be done over the year to spread the cost?

A. Yes, standing order forms will be included with your reminder and can be paid monthly/quarterly/annually.



# VARIEGATE PORPHYRIA (VP)

Variegata porphyria is an autosomal dominant disorder of haeme metabolism and results from defects in the protoporphyrinogen oxidase (PPOX) gene.

The condition is characterised by photosensitivity and propensity to develop acute neurological crises, as well as susceptibility to the acute attack. Skin lesions, fragile skin and blisters are common symptoms.

A study carried out by Dr. J White of Kings College Hospital in London, found that many patients suffer from non-porphyrin skin diseases too. Acne, Eczema, Psoriasis, and Warts were reported by patients in the study. Dr White made several recommendations regarding safe and effective treatments. Dr. Whites' summary of the study advised that sun avoidance and general skin care measures such as the use of moisturizers and sun screens are important in keeping the skin in good condition in those with VP. For skin problems unrelated to porphyria, cream/lotion treatment is generally safe, however table treatments can be unsafe and should be checked with the doctor prescribing the treatment. Monitoring urinary ALA and PBG may be useful.

The standard method for diagnosis of VP is porphyrin analysis of urine and stool. A newer technique is plasma porphyrin fluorescence scanning. Studies at the University of Wales Cardiff, found that in 158 families who have been investigated for VP, 56 different mutations were identified of which 23 appear to be new. DNA screening is now also available for patients and their families when porphyria has been found to be present by other testing methods.

South Africa has the highest incidence of VP in the world due to a founder effect, and over 100 mutations world wide have been reported. Most of the carriers of this genetic deficiency might be asymptomatic throughout their life, however, around a third of them can develop an acute crisis due to the exposure to some triggering agents.

The administration of relatively high amounts of carbohydrates, is the fundamental base of the nutritional treatment, both during the attack and their asymptomatic periods. Carbohydrates act through the mechanism know as "glucose effect".

## BRITISH PORPHYRIA ASSOCIATION GRANT/AID

The trustees and committee of the British Porphyria Association are pleased to announce our new Grant/AID.

Each year we will allocate an amount of money to the Grant/AID fund. Members are invited to apply for Grants/Aid to assist them for various needs or activities, which would improve their quality of life.

Each application should be put in writing to the Trustees/Committee outlining:

1. What is required
2. Why it is required
3. How it will assist in a better quality of life
4. The total cost/copy of invoice
5. The amount of funding required
6. Funding coming from any other agency towards the total cost
7. Your current health status
8. Your current work status
9. Consultants Name and Address
10. Letter authorizing us to contact your Consultant

Each case will be assessed on its own merit. In some cases further information may be requested. If the application is for a project or need that is "time restricted", please indicate this too.

### WHY TODAY?

By Lynda Everest

So much to do so much to say  
Goodness me,  
It's going to be a busy day.  
It's all going wrong I feel very cross  
Who does he think he is?  
He's only the boss.

Why are the kids playing up today?  
Why does no one put anything away?  
That's it I can't cope anymore  
Look at the mess you have made  
on the floor

Unreasonable! Who? Not me!  
Oh leave me alone leave me be.

The world begins to look very grey,  
Negativity takes over pushes good  
thoughts away.  
I feel so tired my limbs a dead weight  
Is that the time? I was sure it was late.

My body feels strange I'm not  
really there.  
Why is everyone looking?  
Do they have to stare?  
The pain increases begins to creep  
Leave me alone, just let me sleep.

I start to vomit, begin to shake  
Did I miss lunch? Not take a break?  
My heart race's sounds like a train  
in my ear.  
Oh this is so unfair;  
I've been well for a year.

At last I no longer have to decide  
or think  
I can't talk, but can hear; silent tears  
make me blink,  
The voices are calm as they  
rush around  
The whirl of the drip a reassuring sound.

I begin to drift float on a cloud  
The train has gone, no pain allowed  
Dream dreams oh so real  
Drift in and out no big deal

Wake up in days totally refreshed  
Crisis over lets get dressed  
So much to do so much to say  
Goodness me it's going to be a  
busy day.

## HELPLINE CONTACT TIMES

Would members please note that our helpline is not manned at specific times, due to work and family commitments.

As a guideline please try to phone during the following hours, when someone is most likely to be in attendance. Outside of these hours the answer machine will be in operation.

**Monday** 19.30 hrs – 21.00 hrs  
**Tuesday** 12.00 hrs – 14.00 hrs  
**Wednesday** Answer machine only  
**Thursday** 14.00 hrs – 15.30 hrs  
**Friday** 10.00 hrs – 14.00 hrs

Weekends and Bank holiday – Answer machine:- Please leave, name, phone number, day and time of message and someone will call you back as soon as possible.

**The BPA Newsletter** is published by the BPA twice yearly.

**Chairman:** John Chamberlayne  
**Treasurer:** Anne Newton  
**Secretary:** Leigh Drake  
**Patron:** Prof. Timothy Cox, Prof. George Elder

14 Mollison Rise, Gravesend, Kent DA12 4QJ  
Tel: 01474 369 231 email: BPA@bodywise.go-plus.net

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

## DONATIONS AND FUND RAISING

We would like to thank the following people for their donations over the past six months.

May Barrington £25 in memory of her uncle, Barbara Price £100 in memory of her aunt, Linda Longman £30 sponsorship for running in the Flora Light, Hyde Park, J Ferguson £10, Heidi Hall £5, Barbara Mather £20, Marion Harrison £15. From a collection at our AGM £24.21 was raised.

Mr & Mrs Fearn, kindly donated £200 to the association at our AGM. Mrs Fearn advised me that they had decided to donate the money instead of having a 70th birthday party for Mr Fearn. The donation will be put into our Grant/Aid fund for the coming year, and we hope that it will help another member. We would like to take this opportunity to wish Mr Fearn a belated Happy Birthday.

Six ladies ran the Flora Light this

year on behalf of the British Porphyria Association. The London Marathon donated £30 back to the BPA from the entry fees paid by the ladies. Well done to all of those who took part, and many thanks.

Once again staff working at Abbey National had a dress down day, with proceeds being donated to the BPA. £170.40 was collected and then matched by Abbey National, making a grand total of £340.80. This is a great way of raising funds, can you arrange a dress down day at your place of work? Why not ask your management. We would be happy to provide you with information and posters about porphyria, so that people taking part have an understanding of the condition. For information how to do this contact our helpline on 01474 369231 or e-mail helpline@porphyria.org.uk.

## Membership/ Donation Form



Registered Charity No.1089609

By making a donation to the British Porphyria Association you will know that you are making a vital contribution 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.

### WAYS TO PAY

Title ..... Name .....

Address .....

..... Postcode .....

Email .....

Telephone .....

Date.....

I would like to pay my annual membership fee of £10

I would like to make a donation of:

£10  £15  £20  £25  £50  other £

I would like to pay by Standing Order (please fill in the form overleaf)

I enclose a cheque made payable to the British Porphyria Association

for £ .....

## STANDING ORDER



Please send this form to: The Treasurers, British Porphyria Association, 11 Blakehill Terrace, Undercliffe, Bradford, West Yorks, BD2 3JS

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) .....

Account no. .... Sort code .....

Starting on\* (date) .....

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

Signed .....

Todays date.....

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

### 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)

Signature .....

Date.....