

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.

New Patron...

At our recent AGM we invited Dr Mike Badminton to be a patron of the BPA.

He was very pleased to accept.

Mike Badminton is head of the porphyria service at Cardiff. As well as seeing porphyria patients at Cardiff, they have a biochemistry lab and offer the only porphyria genetic testing service in the UK. Mike does a great deal for porphyria patients in the UK, as well as co-ordinating links with porphyria experts across Europe.

We are grateful to Mike for all he does for those with porphyria, and are very glad to have him as an additional patron of the BPA.

Full reports of our AGM and conference, and other meetings for the year, will be in our spring newsletter, as there is not room in this one!



Alexandra Rose Day Raffle

This year the Alexandra Rose Day raffle has raised £372 for the BPA.

Many thanks to all those who bought raffle tickets!



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: _____

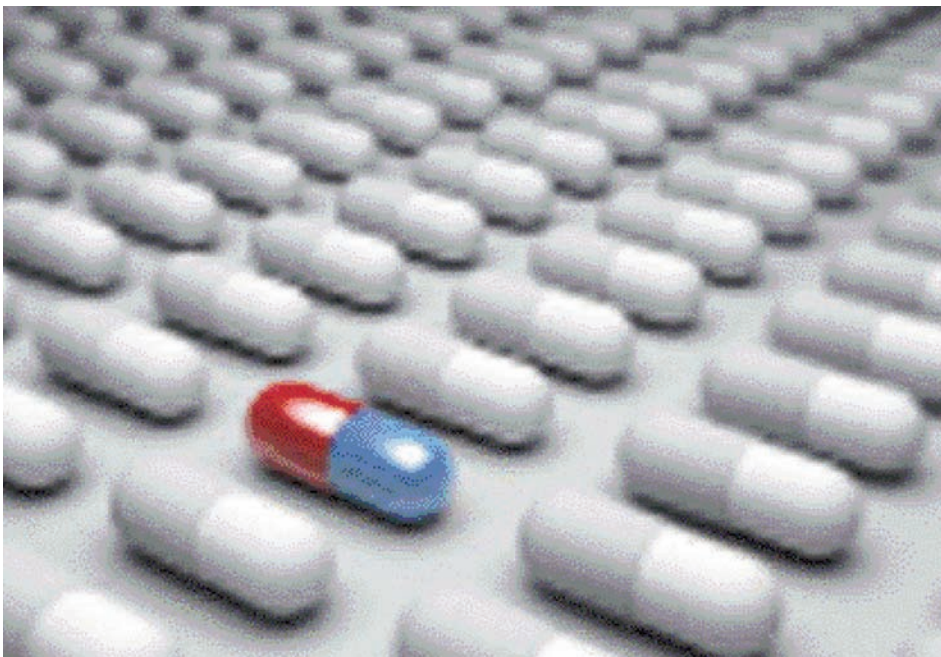
The British Porphyria Association newsletter



Registered Charity No.1089609

ISSUE 15 • NOV 2007

Improving Knowledge of Safe Drugs



The Welsh Medicines Information Centre has been providing information on the use of medicines in porphyria to patients and health care professionals since 1995. However, the amount and quality of information relating to the use of medicines in the acute porphyrias is still somewhat limited. The centre has therefore recently become involved in a three year European public health project (European Porphyria Network: Providing Better Healthcare for Patients and Their Families – Information about Drugs) which aims to improve the quality of information on the safety of medicines for individuals with acute porphyria.

The project involves collecting information, by way of telephone interviews, from individuals with acute porphyria about their experiences with taking medicines safely,

or otherwise, including the development of porphyria symptoms. In obtaining this information it is hoped there will be an improved selection of appropriate medicines to treat various medical conditions in individuals with acute porphyria.

If you:

Have an acute form of porphyria (acute intermittent porphyria, variegate porphyria or hereditary coproporphyria),

and

currently take medication,

and/or

are prescribed a new medication,

and think you may be interested in contributing to this project please contact the project pharmacist, Ms Susan George, on 02920 744861 (answer phone) or email susan.george@cardiffandvale.wales.nhs.uk for further information.

**Contact details for the BPA
are on the back.**

Welcome to “SAKURA Friends”

We received this from our corresponding organisation in Japan in July 2007. 'Japanese porphyria patients group, "SAKURA Friends" (SAKURA means cherry blossoms) was formed in 1997 by porphyria patients, families and support volunteers. Today the group has 66 members.

The group's activities include issuing newsletters three times a year, organizing gatherings and study meetings, providing information and encouraging communication among members in the website, etc.

The group makes efforts to:

- Enhance knowledge about porphyria
- Encourage communication among members
- Support research for early diagnosis and improvement in treatment, which ultimately lead to cure
- Contribute to improve social system and environments for patients and families

We welcome porphyria patients, families and anybody that supports the group's aim and efforts. Welcome!!!

We are also eager to make friends outside Japan so that we can exchange information and experiences and learn from each other.

Those who read Japanese can find more information about our group at www.sakuratomonokai.com



Wanting to come to our meeting, but have difficulty affording the travel?

We have agreed to help members with transport to our meetings if they wish to come and have difficulty with travel costs.

For paid-up members we are willing to pay 15p per mile, or half the receipted train-fare, per family, with a maximum of £50.

You will need to apply in advance, and then we will give you a cheque on the day.

www.porphyrria.org.uk

Antibiotics and hospital acquired infections.

Unfortunately, many bacteria are becoming resistant to antibiotics. Antibiotics may kill most of the bacteria making someone ill, but some resistant bacteria may survive and be passed on. With a resistant strain, like MRSA or *Clostridium difficile*, few antibiotics can kill them. This can be a particular headache if the patient has an acute porphyria, limiting the choice of antibiotics even further. It may mean having to risk an acute attack (which can be treated) to control the infection.

Responsible use of antibiotics is important if those we have are to remain useful. Developing new ones can take a long time.

What can we do to help?

- Don't expect your GP to give you antibiotics if you have a viral illness, like a cold. Antibiotics do not kill viruses.
- If you are given antibiotics, take the whole course, even when you start to feel better. This will make sure that as many bacteria as possible are killed off, even those with some resistance.
- When you visit someone in hospital, wash your hands before and after visiting. Most wards now have a wall dispenser by the entrance which gives alcohol rub, so you can use it going in and going out.
- Try not to wear dirty clothes when visiting. Taking an infection to a sick person could be disastrous, as could taking one home to family and friends. Many hospitals have changed dress codes for staff recently – short sleeves, no uniforms outside the hospital and no ties for the men. It all helps reduce infection.
- If you have to go into hospital yourself, take a few simple precautions. Use your own toiletries (soap, towel, etc.). Baby wipes, or antiseptic wipes, can be useful if you are likely to be stuck in bed. Clean your hands after using the toilet or a bedpan, and before you eat. Wear slippers or shoes out of bed to keep your feet clean. Tell the nurse in charge if your locker, table or floor needs cleaning.
- If a doctor or nurse needs to examine you or give you an injection, don't be afraid to ask whether they have washed their hands. If you have had surgery and a dressing needs changing, staff should wear gloves and an apron.

Porphyria Interest Group

This year, the Porphyria Interest Group met in Dublin, in September. This is a group of medical and laboratory staff who have an interest in the diagnosis and treatment of porphyria.

Our chairman, John, gave a talk on the results of the questionnaire we sent out to our members, on the acute porphyrias. It provoked some discussion, particularly on how to judge whether someone was having a mild attack.

There were talks on EPNET, the EU funded project. The porphyria-europe website is now available in a number of languages, and information on non-acute porphyrias is progressing. Unfortunately, getting a consensus on treatment can be a lengthy process as everyone has their own ideas! The Welsh Medicines Information Centre is one of four currently looking at patient experience with medicines (see separate article). Kings College Hospital is one of four centres looking at quality assurance in testing, to try to establish standard methods and find sources of test samples, so accuracy can be checked. There is also a group working on a study of new cases over 3 years, covering long-term implications for health.

Another talk looked at the problems for porphyria reference labs. Most samples sent for PBG testing are to check whether the patient is having an acute attack, but some routine monitoring can also be useful. The problem of standard methods was raised again. Some labs weigh faecal samples wet, others weigh them dry!

A curious case of PCT was described. This was a female patient who had breast cancer, and a drug used for this cured the PCT fast – it stops the production of oestrogen (one of the female hormones). Drugs can often have more than one effect.

King George III's Malady: Re-evaluation of His Porphyria Diagnosis

Professor T. J. Peters, Ironbridge Institute, University of Birmingham.

In 1966 Drs. Macalpine and Hunter stated, on the basis of their examination of the medical records of George III, that he suffered from acute intermittent porphyria. In 1968 on the basis of studies of his ancestors they changed the diagnosis to one of variegate porphyria and claimed this was The Royal Malady. In spite of well-argued objections by acknowledged experts they persuaded historians and not a few medical doctors that their case was proven.

My careful re-examination of his medical records, in collaboration with Dr. D. Wilkinson, a professional historian and expert on the life and times of George III: Willis papers (52 vols. British Library) and Manners Sutton Papers (32 vols. Lambeth Palace Library) and the Dairies of various Court Officials, indicated that Macalpine and Hunter were highly selective in their presentation of the data and that their conclusions were flawed. Thus, they report discoloured urine on four occasions over the 30-year period of his recurrent illnesses but fail to mention five occasions when, in the six weeks immediately prior to the so-called 'blue urine': his urine was of normal colour. Three days prior to the 'blue urine' he was commenced on pills containing extract of gentian, a material quite capable of causing the blue discoloration!

The King had episodes of obstructive jaundice with yellow-brown urine and pale stools consistent with a diagnosis of cholelithiasis and diagnosed as gall stones by his attendant physician Sir George Baker. Similarly, Macalpine and Hunter claimed that his transient lameness, senile cataracts and episodic hoarseness were characteristic of variegate porphyria when it is clear that there are other much more likely explanations apparent in the notes.

The reason for the persistent misinterpretation of the data and their uncompromising claims has been explored. Macalpine and Hunter clearly at the time had an agenda that all psychiatric disorders were medico-biochemical in nature and this belief also pervaded their clinical practice. In labelling the King's illness a metabolic disorder they hoped to reinforce their agenda. The acceptance of the porphyric diagnosis by historians both professional and populist has occurred because they themselves failed to review the King's primary medical records.

Careful review of the King's symptoms supports the earlier reports of manic depressive psychosis in that the King had Type I bipolar affective disorder with mood congruent delusions and an Alzheimer-type senile dementia in the last decade of his life before his death aged 81 years.

Our conclusions are that the studies of Macalpine & Hunter of the King's malady were seriously flawed and thus any derivative studies should be viewed with similar suspicion. They had an agenda that psychiatric disease was metabolic in nature and the porphyric diagnosis was pursued to support this aim. Re-evaluation of the King's illnesses support earlier diagnoses of cholelithiasis with bipolar affective disorder complicated by a senile dementia.



PORPHYRIA TESTING AT BIRTH FROM UMBILICAL BLOOD

In May this year the laboratories at Cardiff received samples of blood from the umbilical cord of a baby born to a member of the British Porphyria Association.

DNA tests confirmed that the baby had inherited Variegate Porphyria from her father. Surprisingly the blood plasma scan showed that porphyrin levels were unexpectedly high at the time of birth.

This revelation has left us with some interesting questions.

1. Do all those who inherit an acute porphyria have raised porphyrins at birth?
2. What causes the raised porphyrins, could it be the stress of being born, or perhaps hormones released at the time of birth by the mother?
3. Will the levels subside over the first few months to those expected of a baby?
4. Could long term studies be used to predict who may or may not go on to develop acute symptoms?

Dr. Mike Badminton, one of our medical advisors, reports that another porphyria group had seen a positive scan from cord blood, however, genetic testing (DNA) was negative and the plasma scan returned to normal. In this case the mother was the affected parent and it was therefore assumed that this was due to a transfer of porphyrins across the placenta.

In the long term this may be an area of research that the BPA can look to help fund. In the short term, it would be helpful for the Cardiff team to receive more umbilical blood from babies of known patients to help increase knowledge and understanding.

If you are a porphyria patient expecting to become a parent you may like to consider having your baby tested at birth. This can be arranged via clinical genetic services.

It is a painless procedure, as blood is taken from the umbilical cord and the baby does not have to have blood drawn from a vein.

You will need to consider the implications of testing at birth, and it may be wise to speak to a genetic counsellor before you make a decision.

Dr Badminton stresses that it is important that anyone who has tested positive for porphyria should inform their relatives and advise them to be tested too, so that future generations are aware and can make informed decisions about their lifestyle.



FUNDRAISING

Many thanks to all those helping us to raise money: Karen Harris and her husband, Graham, ran the Great South Run in October and expect to raise over £400 in sponsorships. Despite driving rain and wind they managed to complete the 10 mile course in 1 hour 33 minutes. A Summer Ball held in memory of Helen Gibbs raised a fantastic £2,500, again, many thanks to all those participating.

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 £400 for The British Porphyria Association just by shopping through www.buy.at/BPA.

Retailer	Commission	
An ISA from Legal & General	£85	
Sky HD package	£110	
A contract phone from Dial-a-phone	£60	
Home insurance from the CO-OP	£70	
Powergen dual fuel	£35	
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	£422.50	