Issue 37 / **December 2018**

THE
BRITISH PORPHYRIA ASSOCIATION
NEWSLETTER

Registered Charity No. 1089609

SCENESSE® PROGRESS

The NICE (National Institute for Health and Care Excellence) process for the approval of Scenesse for use on the NHS in England has been dragging on for some time now, with no clear end in sight. Similar processes in Scotland and Wales are currently on hold.

Following a December 2017 recommendation not to approve Afamelanotide (Scenesse), an appeal hearing was held on 30th July 2018 – which the BPA, the British Association of Dermatologists (BAD) and the International Porphyria Patient Network (IPPN) attended. The outcome of the appeal hearing was published in October.

In summary, the appeal panel upheld some of the stakeholders' appeal points: (1) that NICE failed to include the international patient group in a committee meeting, (2) that NICE used assessment methodologies likely to have discriminated against EPP patients and (3) that it was unreasonable of NICE to state that the trial results showed small benefits of the drug.

Since then, the BPA and other stakeholders involved have been invited to submit any new or additional information or evidence by the middle of January. This will then be assessed in a NICE committee meeting in March 2019.

So what does this mean to UK patients?
Unfortunately, this is still unclear. Whether or not this decision will lead to a change in the recommendation remains to be seen. What we can say at the moment, is that it is no longer a definite no.

The BPA will continue to work with the other stakeholders to put the case forward for those with EPP.

Why is the process with NICE so lengthy?
With a limited pot of money for NHS health care, there are difficult choices to be made regarding how best to spend it. NICE is the independent body which makes these tough decisions. If NICE recommends a new expensive treatment for a particular group of patients, something else has to give. Turning it down, however, denies a treatment to patients who might have benefited from it. This means there are tricky decisions to make between, for example, prioritising a treatment that might save or prolong the lives of a few people, an asthma or diabetes intervention that could help millions, or a drug that could improve the quality of life for a certain group of people?

With such a responsibility, there are lots of different stages in the process and huge amounts of data and information to obtain from different stakeholders. In the case for Scenesse, the usual timescales have been extended even further to obtain more evidence.

Further details can be found on the NICE website at: *www.nice.org.uk/guidance/indevelopment/gid-hst10009/*

GDPR OPT IN REMINDER

Recent changes to data protection laws give you more control over your personal data, including why data is held by organisations and how you can be contacted. If you still wish to receive our newsletters, leaflets and information on our events, we now need your explicit consent.

Many thanks if you have already updated your contact details and confirmed what information you would like to receive from us, and how.

For those who haven't yet done this, please go to *www.porphyria.org.uk* and select 'Opt In' to complete the short online form. Alternatively, please call 0300 30 200 30 to consent by phone.

ACUTE PORPHYRIA STUDY

The BPA, along with Alnylam Pharmaceuticals and BresMed (an independent health economic and outcome research consultancy), is conducting an online survey on how acute porphyria impacts the lives of affected individuals and their carers. The anonymised results of the survey may help explain the burden of acute porphyria to health authorities and the public. To participate in the survey, please visit the BPA website *www.porphyria.org.uk*.

ACT NOW: BOTH STUDIES CLOSE IN JANUARY

EPP STUDY

The Photobiology team at Salford Royal Hospital are seeking adults with EPP to take part in a study examining the relationship between daylight exposure and occurrence of symptoms throughout the year. The study, funded by the British Skin Foundation, is recruiting now. Participants will complete a diary for up to 4 weeks in each season to record their time spent outdoors, weather conditions, clothing and sunscreen worn, and occurrence of symptoms. Attendance at the hospital is not required. For further information, please contact Joanne Osman, Senior Research Nurse on 0161 206 0457; email: *joanne.osman@manchester.ac.uk*.

FUNDRAISING AND AWARENESS

There are lots of fundraising events being planned for 2019. **Active events:** if you would like to fundraise for the BPA through an active event, there are two main ways of doing it.

You can register for one of our official charity places:

- Ride London-Surrey 100 (Sunday 4 August 2019): 2 places
- Great North Run (Sunday 8 September 2019): 5 places: fundraising target £300

Places are allocated on a first come, first served basis, so please let us know ASAP if you or someone you know is interested on *liz.gill@porphyria.org.uk* and we'll send you more information.

Or you can obtain a public ballot place and still raise funds for the BPA:
If you use your own ballot place to run/cycle for the BPA, we don't have to spend anything on your place. There's no fundraising target for you and however much you raise, your contribution will make a massive difference to porphyria patients.

Some of the popular public ballot deadlines are shown below, but you might find another event that you would like to do instead. If you are lucky enough to be successful in a ballot and want to raise funds for us, please get in touch to see how we can help.

London Marathon 2019: already closed. London Marathon 2020: not set yet, but the date is likely to be around the last week in April or first week in May 2019. It always has a short entry window, so keep an eye on the following page for more details: *www.virginmoneylondonmarathon.com/en-gb/how-to-enter/*

Great North Run 2019: The ballot will open in January and you can sign up to their reminder service *http://support.greatrun.org/reminderservice/?id=881* to be the first to know when you can register.

The 2019 Prudential RideLondon-Surrey 100: The ballot closes at 17:00 on Friday 4 January 2019 or when 80,000 registrations have been received, whichever is earlier.

International Porphyria Awareness Week (IPAW) 2019
Alternatively, you might want to do something completely different. How about trying our coordinated IPAW 2019 ideas, where we hope as many of you as possible can do something easy to raise awareness. IPAW will start on the third Saturday in April and runs for 8 days, so for 2019 it is the 20 – 27 April.

- Bake sale: we can provide BPA logo cake toppers for your cakes or buns
- Bag packing event: contact us for a step-by-step guide on how to organise a bag-pack event at your local supermarket.

SKIN PROTECTION AND DUNDEE CREAM

It has come to our attention that people in England have been experiencing problems obtaining Dundee Cream on prescription since the NHS stressed that sunscreens should not be routinely prescribed. Dundee Cream is different to a normal sunscreen, as it is a large-particle-size reflectant sunscreen, so it should still be available. However, pharmacists in some areas report that they can no longer select sunscreen options on their systems and patients are referred back to their GP.

GP practices in some cases have to refer to a committee to be able to prescribe it.

There are definite misunderstandings about the process:

- A prescribed sunscreen for severe photosensitivity, as for example in EPP, is allowed and nothing in the English NHS guidance has changed this.
- In the skin porphyrias, the problem is with visible light NOT ultraviolet which is why normal commercial sunscreens do not help much.

Specialists and Tayside Pharmaceuticals from Dundee are writing a letter, hoping that when this is published it could be referred to if someone is having difficulty obtaining Dundee Cream.

Unfortunately, people cannot simply purchase Dundee cream as it is a medicinal product, rather than a cosmetic, and can only be obtained with a prescription.

While this is problematic, our advice is to be persistent. You may need to ask your GP to refer the case to a committee to enable it to be provided.

The following document might be useful in trying to persuade GPs/ CCGs to prescribe:

- The British Association of Dermatology list of recommended ‘Specials’ (page 11) states that reflectant sunscreens are recommended for photosensitivity disorders *www.bad.org.uk/shared/get-file.ashx?itemtype=document&id=1848*

In the meantime, mineral sunscreens containing zinc oxide and/or titanium dioxide offer a level of protection by providing a physical block to visible light. Generally, the larger the particle size, the greater the protection. So the creams that offer the greatest protection are likely to be thicker, messier and quite opaque (like Dundee Cream).

Our members have noticed some success with the following, although this is all trial and error and you may not find the same products work for you.

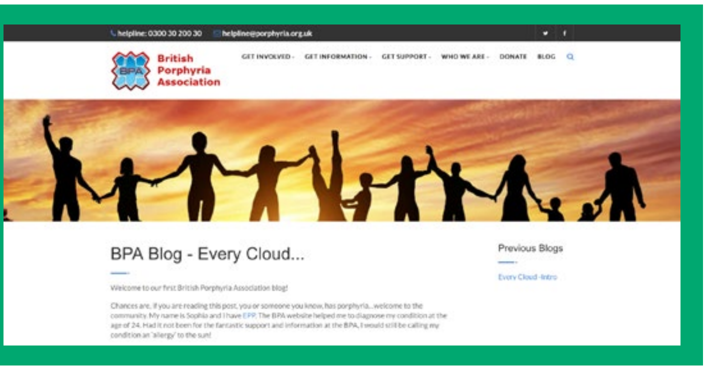
- Surf Mud natural zinc
- Invisible Zinc Broad Spectrum 50 SPF
- Neutrogena Sheer Zinc-Oxide Face dry-touch sunscreen 50 SPF

Dr Robert Dawe recently suggested that infrared protection cream might be useful to people with skin porphyrias due to the long wavelength that they target (even longer than visible light), and believes that this area requires further research. So although not confirmed, if you are happy to experiment with new things, it might be worth exploring this avenue with your specialist or dermatologist, especially in the absence of other availability.

We’d be interested to hear how you get along.

BPA BLOG!

We’re really excited to announce that the new BPA blog is now live. Our newest committee member, Sophia, who has EPP, was eager to create a new source of community for all the porphyrias. Sophia aims to provide fun and useful tips and advice on living with porphyria, as well as focusing on the mental and emotional wellbeing of living with a long-term health condition. This brilliant new feature can be found NOW on *www.porphyria.org.uk.*



BPA Blog - Every Cloud...

Welcome to our first British Porphyria Association blog!

Chances are, if you are reading this post, you or someone you know, has porphyria...welcome to the community. My name is Sophia and I have EPP. The BPA website helped me to diagnose my condition at the age of 24. Had it not been for the fantastic support and information at the BPA, I would still be calling my condition an 'allergy' to the sun!

THANK YOU

This is our spot to extend huge thanks to all of the people who take the time and effort to raise funds and awareness for the BPA. We are hugely grateful for all the support that you and your family/friends provide. Without you, we wouldn't be able to support so many people through their porphyria journey. Here's just a selection of our wonderful fundraisers in the last six months.

Gillian Chapman and her family and friends have been busy with lots of events. They sold merchandise at the Valerie Shepherd Dance Academy, bringing in £97, whilst also running a Jeans for Genes day, which raised funds for the BPA, as well as also donating to other rare disease charities. Mr & Mrs Irwin's gold wedding anniversary celebration in August led to £315 being donated to the BPA and after hearing about Matthew and Sarah's lives with EPP, a family member, Mr Brian Crammon, kindly donated a huge £1,000 to the BPA.

Rachel Harrison ran the Henley Triathlon and raised a wonderful £100. While Jean Barrett at the Welcome Café also raised a significant £100. Thank you to you all.

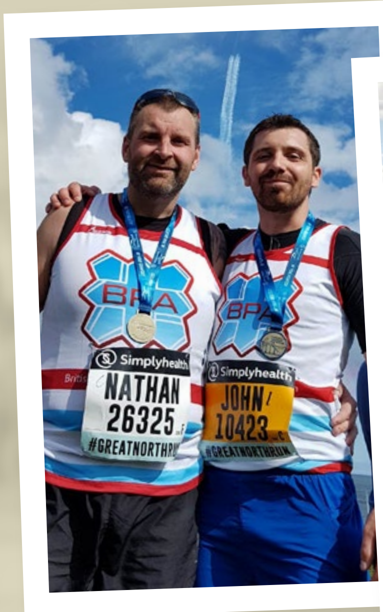
The BPA, and specifically Sue Burrell, would like to extend a huge thank you to John Critchley, the founder of Integrated Technical and Consultancy Services for his very kind donation of a top-of-the-range laptop, which was fully set-up ready for Sue and the BPA to start using at the Conference in Reading in October. It is expected that this donation will have equated to around £1,600 – a massive and very well received contribution to the BPA. This will have not only a huge impact on Sue's ability to work responsively, but it will also enhance technical capabilities at meetings and events in the future.

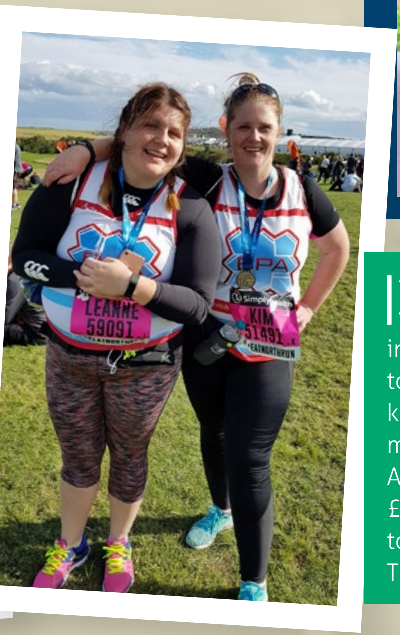
GREAT NORTH RUN AND GREAT SOUTH RUN 2018

Our team of nine Great North Run runners this year battled a hot September day to all complete the race. Nathan Abbott, Ryan Arrowsmith, Cath Clough, John Critchley, Kim Dickson, Leanne Geritz, Rebecca Jellis, Becki Pedelty and Sarah Wall, formed the GNR team.

In October, Jamie Wentzell, Daniel Wentzell, Karran Speakman and Natassja Chadwick represented the BPA at the Great South Run.

Both teams together raised an amazing, target-smashing £4,498. Huge THANKS to them and all their networks who donated and supported them to make them really memorable events.





SHADOW HOPPING

Natassja Chadwick, mum of two children with EPP created a Shadow Hopping Day and organised an awareness day at Tesco. The team raised a whopping £1,115. As a further surprise, Natassja's husband managed to obtain an outstanding £1,000 donation from his workplace. To complete her challenge, Natassja and her friend Karran took part in the Great South Run too. In addition, Natassja managed to create loads of publicity through an interview on Manx Radio and an article in the local newspaper.

You can listen to the interview here: *www.facebook.com/ManxRadio/videos/410433519483597/*

Well done and thank you for such a successful and thorough campaign.

It's also great to see young people being involved in fundraising too. Amelia Bell, age 8, knows how much her mum has suffered with AIP and raised a fantastic £131 by shaving her head to raise funds for the BPA. Thank you Amelia.

EVENTS 2018

The BPA held two events this year, one in Glasgow and one in Reading. We were delighted to see so many new faces with a record number of 124 people attending in total. The feedback was very positive and we look forward to building this into next years' conferences.

We'd also like to extend a sincere thank you to Orphan Europe, who again donated a significant amount, through a grant, to help us put on both of these amazing events.

BPA EVENTS 2019

We are delighted to be able to invite you to two BPA events in 2019.

IRELAND CONFERENCE – SUNDAY 23 JUNE 2019

We are very excited to be holding our very first conference in Ireland, this will be at the Ashling Hotel in Dublin city centre. The invitation is open to all, but priority will be given to those members based in Ireland or those who might not have been able to get to other events on the mainland.

The programme is yet to be finalised but, as usual, there will be doctor talks with the support of various porphyria specialists from across the UK and Ireland, as well as patient talks, updates on research and plenty of opportunities to speak with other porphyria patients and their families.

NORTH-WEST, ENGLAND – PORPHYRIA EVENT – SATURDAY 12 AND SUNDAY 13 OCTOBER 2019

We are delighted to announce that to celebrate the 20th anniversary of the BPA forming, we will be holding an exciting new event with a completely different format. This will take place in the Greater Manchester area, over the weekend of 12 and 13 October 2019. It will be much more family oriented, with lots of fun elements for all the family. We will still have doctor and patient presentations, but they'll be presented with a twist and you'll have the opportunity to only attend the parts that interest you!

More details will be circulated in due course. Keep a look out, as we expect this event to be extremely popular.

RESERVE YOUR PLACES

We very much hope to see you at one of these exciting events, both events are free to attend, but registration is essential as spaces are limited. If you would be interested in attending, please forward the names of anyone wishing to come with you to *sue.burrell@porphyria.org.uk* or call 0300 30 200 30.

Travel bursary: The BPA have funds to help support our members to attend meetings. If you would like to apply for funding, please get in touch (funding available up to: £50 for travel and £50 for hotels).

INTERNATIONAL CONGRESS ON PORPHYRINS AND PORPHYRIAS (ICPP)

The two-yearly ICPP is being held in Milan in September 2019. Before the main scientific conference begins, a patient day will take place on Sunday 8 September. Organised by patient associations, it is a fantastic chance for international patients and organisations to communicate with and hear from specialists and other patients.

A scientific programme will then take place over the following three days. During this time, the latest research developments in the field of porphyria will be presented and discussed. The BPA will attend this conference, so that we can convey the information back to our members.

HELPLINE

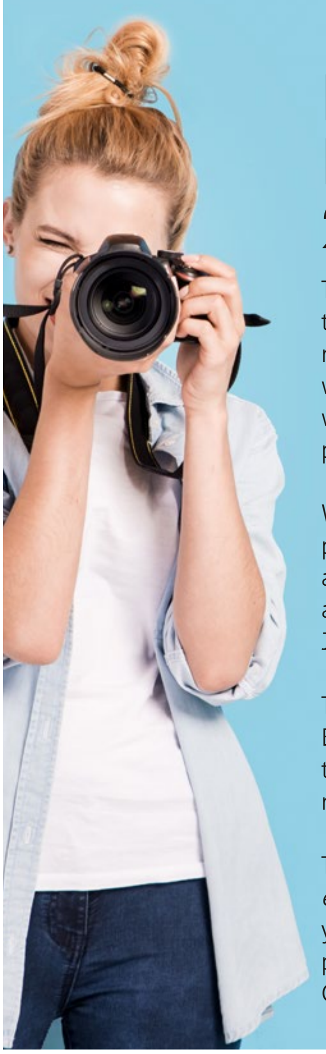
0300 30 200 30

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

web: *www.porphyria.org.uk*
email: *helpline@porphyria.org.uk*
address: BPA, 136 Devonshire Rd
Durham City, DH1 2BL



EURORDIS PHOTO AWARD 2019

The EURORDIS Photo Award is an opportunity to visually express what it means to live with a rare condition. Open to everybody around the world, photos can reflect any element of living with a rare condition – especially the resilience patients have to adopt in the face of adversity.

World-renowned National Geographic photojournalist Marcus Bleasdale will select a shortlist of 5 photos that will be opened to a public vote. Online voting will start on 15 January 2019.

The winner will be announced live at the EURORDIS Black Pearl Awards on 12 February, the three finalists with the most votes will each receive a prize. 1st prize is an iPad.

To enter, visit *https://blackpearl.eurordis.org/eurordis-photo-award-2019/* and upload your photo before 7 January 2019. Once a photo has been approved, it will appear in the Gallery.

THE **BRITISH PORPHYRIA ASSOCIATION** Registered Charity No. 1089609

MEMBERSHIP / DONATION FORM

MEMBER DETAILS

Please complete (*required field) and return to:
BPA Treasurer, Rothlea House, 7-8 Quarry Lane, Butterknowle, Bishop Auckland, DL13 5LL or email it to treasurer@porphyria.org.uk

Title * Full name *

Address *

Postcode *

Landline Mobile

Email

Type of porphyria

Are you a (please tick):

☐ Patient ☐ Relative ☐ Medical professional ☐ Supporter/donor ☐ Other

What information are you happy to receive? Tick all boxes that apply:

☐ All BPA communications ☐ Newsletters
☐ Patient events/information ☐ Fundraising/awareness events

How would you like us to keep in touch with you? Tick all boxes that apply:

☐ Post ☐ Email ☐ Telephone ☐ Text message

DONATIONS

By making a donation to the British Porphyria Association you will know that you are making a vital contribution to our work. A suggested donation of £15 per year will help us raise awareness, man our helplines or support research.

I would like to make a donation of:

☐ £10 ☐ £15 ☐ £20 ☐ £25 ☐ £50 ☐ other £................................

WAYS TO DONATE:

☐ Cheque: made payable to the British Porphyria Association.

☐ Online banking (one-off payment or regular standing order):
BPA bank account: Sort code: 20-43-63, Account no. 7099 6904
Please use your name and postcode as the payment reference.

☐ Credit card: via Just Giving (*www.justgiving.com/britishporphyriaassoc*).

☐ If you would prefer to complete a standing order form, please call us on 0300 30 200 30 and we will arrange to send one to you.

☐ I would like a receipt

DO YOU PAY UK TAX?

If you pay UK tax, the BPA can reclaim 25p of tax on every £1 you give. This does not cost you anything and does not affect your personal tax position. Simply sign and date the Gift Aid declaration.

GIFT AID DECLARATION

I am a UK taxpayer and would like the British Porphyria Association (Registered Charity No.1089609) to treat all donations that I have made in the last six years and all future donations that I make from the date of this declaration as Gift Aid donations. I understand I must pay an amount of income tax and/or capital gains tax equal to the tax reclaimed on my donations. (I will advise the BPA if my tax status, name or address changes.)

DATE SIGNATURE