

THE BRITISH PORPHYRIA ASSOCIATION NEWSLETTER

Registered Charity No. 1089609



GLOBAL PORPHYRIA DAY

WED 19 APRIL 2023

The BPA engaged with the global porphyria community on Wednesday 19 April 2023 for Global Porphyria Day (GPD). Previously known as Porphyria Awareness Week, this wonderful global event provided patients (and their families and friends) with the opportunity to raise awareness about porphyria as well as opportunities to get involved in the theme of **This Is My Porphyria**.

The global community will be releasing a series of porphyria type specific videos over the coming months. Launched on Global Porphyria Day, with an introduction to the porphyrias and some of the amazing patients involved in sharing their stories, this highlighted the start of the **This Is My Porphyria** awareness campaign.

The first **#MyPorphyria** video can be found on

YouTube: <https://youtu.be/QsGt4upaJYw>, you can search for others via the social media tag **@GlobalPorphyria**

It's not too late to get involved

Check out our social media channels for ideas on how you can still get involved, even after the main day in April:

- **Purple For Porphyria:** Get your work/school/ clubs involved in #PurpleForPorphyria, why not 'wear purple' or do a 'purple bake-sale' and raise funds and awareness of porphyria in your community!
- **Share your story,** or re-share stories/videos from our social media channels to engage with #MyPorphyria to tell people how you are affected by showing This Is #MyPorphyria!
- Take the opportunity to help **raise awareness** of



This is
#MYPORPHYRIA
GLOBAL PORPHYRIA DAY

porphyria throughout 2023 using: **#MyPorphyria** **#PurpleForPorphyria** **#Porphyria** **#GPD2023**

If you'd like to share your story or want support from the BPA to get involved, please email us helpline@porphyria.org.uk



CAN YOU HELP THE BPA?

We are looking for general and specific expertise that could help to support the BPA, on an ad-hoc and as needed basis, in areas such as fund generation and fundraising; general accounting and finance; and, legal advice.

Does your company allow you to contribute 1 or 2 days voluntarily to help a charity? Or, could your company offer a little pro bono time to help the BPA? If so, and you have some specific skills/ experience, please do get in touch: helpline@porphyria.org.uk

EVENTS 2023

RECONNECT SNOWDOME CONFERENCE SATURDAY 3 AND SUNDAY 4 JUNE 2023

Snowdome Conference Facility
in Tamworth (near Birmingham),
Staffordshire, B79 7ND

Day 1: 16:00-20:00 Social evening:

All welcome. This is a chance
to CONNECT with others in the
porphyria community and get
involved in some social activities –
refreshments provided.

Day 2: 10:00-15:30 Patient day:

All welcome. The day will include
doctor and patient talks, updates on
research and plenty of opportunities
to speak with other patients.

Register now: www.eventbrite.co.uk/e/bpa-reconnect-tamworth-3-4-june-2023-tickets-596749552977

SAVE THE DATE:

RECONNECT ALFRESCO SATURDAY 23 AND SUNDAY 24 SEPTEMBER

Following the success of our
activity day in October last year,
we are delighted to be developing a
two-day residential which will take
place in Thetford Forest, Norfolk. The
event is being designed primarily to
offer a safe residential environment
with numerous outdoor activities for
children and young people affected
by erythropoietic protoporphyria
(EPP), but all are welcome to get
involved in a number of outdoor
active challenges!

For more details on the venue
and location, check out Thorpe
Woodlands website: <https://www.thorpewoodlands.co.uk/>

The venue is situated in a lovely
sheltered forest providing excellent
tree coverage from the sun. There
are numerous activities on offer and
flexible accommodation – the main
building has excellent dorms for
those aged 7 and up, and there are
also some pods, which we hope to
be able to reserve for young adults
and for any families wishing to
attend/stay with younger children.
We are currently finalising details on
how the day will work and looking
into funding options, but we have
approximate timings of: Arrival 10:30

Saturday 23 September 2023, with
a departure of early afternoon on
Sunday 24 September 2023.

We understand that this type of
activity may feel unreachable for
EPP patients, but the adaptable
venue and activity options available
at Thorpe Woodlands will be perfect
for those with EPP. To help with
the process, we will be offering for
non-EPP siblings or friends to come
along, to stay with their friend/
family member in order to help
support them. This approach should
help those attending, of all ages,
but it will also allow for parents/
family members to have plenty of
opportunities to socialise with other
parents of children/young people
with EPP.

Please get in touch ASAP to
register your interest as we expect
demand to be high and places
will be limited. Please contact
helpline@porphyria.org.uk to show
your interest and we will then
arrange a screening call with you to
discuss the next steps.

We are extremely excited by this
project and look forward to reserving
your places at this amazing BPA
Alfresco Residential event.

VACANCY:

Volunteer Treasurer/Trustee

We are looking for an enthusiastic and
organised candidate to take on the role
of Treasurer for the BPA.

The role of Treasurer is of vital importance
all of the time, but we are also seeking to
grow the organisation and are looking for the
right person to help us achieve our strategic
aims to reach more people affected by
porphyria, and especially young people.

A competent treasurer or part-qualified
book keeper or accountant would be
ideal. Experience in the charity field is
desirable but not essential. Our accounts
are not complex, but we do have multiple
income streams to consider. The successful
candidate would need to maintain financial
control and ensure we comply with our legal
responsibilities as a Registered Charity.

This role provides an excellent opportunity
for developing your experiences and would
be seen favourably on your CV. In terms
of time commitment, we expect the role to
take around 2 hours a week, as much of the
input and admin will be done by our new
assistants.

Find the full details at www.porphyria.org.uk/vacancies

Applications must be made via Liz Gill,
Strategic Lead of Operations on liz.gill@porphyria.org.uk and copy to helpline@porphyria.org.uk. Please provide your CV,
along with a covering letter stating why
you wish to work for the organisation, and
demonstrating how your skills meet the
essential criteria and would add value to the
Trustee Board.

We're looking for... VOLUNTEER TREASURER

Could you help us grow?

Requirements

- Financial experience
- Computer literacy
- Good communication skills
- Able to dedicate 1-2 hours per week



www.porphyria.org.uk

THE
BRITISH PORPHYRIA
ASSOCIATION

SCENESSE® NOT RECOMMENDED FOR USE ON THE NHS IN ENGLAND

The BPA is very disappointed with the recent release of the NICE Final Evaluation Determination (FED) regarding SCENESSE® (afamelanotide) which is deeply frustrating for EPP (erythropoietic protoporphyria) patients.

The technology appraisal process for the use of SCENESSE via the NHS in England has been ongoing for more than 5 years. The decision by the NICE committee 'not to recommend' leaves EPP patients in an incredibly difficult position with no effective treatment available on the NHS to improve their condition.

Why is the outcome 'not to recommend'?

The outcome of this final committee meeting depended on certain requirements being met and/or more data being gathered to reduce uncertainties about the effectiveness of the medication. So far, this has not been achieved, therefore NICE has been unable to recommend the medication via the usual routes, or via a Managed Access Agreement (while more data is gathered).

What happens next?

Stakeholders can appeal the decision if they feel that 'NICE failed to act fairly or exceeded its powers', or 'the recommendation is unreasonable in the light of the evidence submitted'.

Regrettably, the BPA does not intend to appeal this final decision! This appraisal has taken a huge investment of BPA resources, where we have collaborated with NICE, Clinuvel, the international community and clinicians through four

separate rounds of consultations and an appeal to put forward the patient perspective. Despite significant sustained effort and energy, maintained since the process began in 2017, we feel the optimal strategy is to allow the company to appeal NICE's decision through whatever channel they feel most appropriate. We remain engaged in the wider process and available to advise and support Clinuvel, NICE and wider stakeholders in the best interest of our patients. This route maximises the BPA's capability to support porphyria patients and their families.

In the longer term, NICE remain open to the submission of further evidence that improves their assessment of cost/benefit of the treatment to bring it within the thresholds that they apply for treatments provided by the NHS.

On the positive side...

While the outcome of the appraisal is negative, the BPA is pleased to see that the final evaluation document demonstrates an improved understanding of EPP. The final assessment also revealed a significant closing of the gap towards meeting the cost/benefit thresholds NICE applies when assessing treatments for prescription via the NHS. Largely as result of the combined efforts of international patient groups,

working in collaboration with the British Association of Dermatologists (BAD) and other clinical experts, there has been a step-change in demonstrating the impact that the condition has on patients and families compared to the way it was perceived at the beginning of the process. A hollow victory in the face of a negative outcome, but a success that will help in future appraisals.

Further, whilst the outcome is disappointing, we are sure this will not be the last we hear from Clinuvel.

Specific Recognitions

The BPA would like to take this opportunity to thank all that have been involved in the process to date, especially Prof Lesley Rhodes and Dr Bob Sarkney. In particular, tremendous thanks are due to EPP patient, expert and scientific researcher Dr Jasmin Barman-Aksözen, and her colleagues within the IPPN (International Porphyria Patient Network), who continues to work tirelessly in developing studies and evidence related to EPP and who has been an influential stakeholder in the process, alongside the Global Porphyria Advocacy Coalition (GPAC) too.

If you would like to stay informed about future developments for EPP, please register as a member of the BPA via our *opt in* form.

Register now

Patient Day

Snowdome, Tamworth

near Birmingham

Saturday, 3 June 2023 16:00-20:00
Sunday, 4 June 2023 10:00-15:30

 



UKPMIS

UK Porphyria Medicines
Information Service

DON'T SUFFER THIS SPRING – HAY FEVER AND PORPHYRIA

An update from UK Porphyria Medicines Information Service

Spring is finally appearing but this does mean that the pollen count is rising. The rising pollen count can cause symptoms of hay fever to appear. These symptoms often include sneezing, a runny nose and itchy eyes.

Hay fever remedies

There is no cure for hay fever but there are some things you can do to help reduce the amount of pollen you come into contact with:

- put Vaseline around your nostrils to trap pollen
- wear sunglasses when outside
- shower and change your clothes after going outside
- stay indoors whenever possible
- keep windows and doors shut
- vacuum regularly and dust with a damp cloth

If these don't help, there are medications that you can try, including:

- antihistamines
- eye drops
- steroid nasal sprays

A pharmacist will be able to guide you on how to use and take the medications. Make sure you talk to your pharmacist or doctor before starting new medications.

Antihistamines

Antihistamine medication is available as either a tablet, liquid, nasal spray or eye drops. Antihistamines help to reduce symptoms of hay fever within an hour of taking them. Commonly used antihistamines that are safe for people with acute porphyria include:

- cetirizine
- fexofenadine
- loratadine

Older antihistamines make some people drowsy. These may be used if symptoms are affecting

how well you sleep. Chlorphenamine is an older antihistamine that is safe for those with acute porphyria.

Eye drops

If you get painful or itchy eyes as a symptom of hay fever, you can try using eye drops as well as the above antihistamine medication. There are different types of eye drops available that are safe for those with acute porphyria including:

- azelastine
- sodium cromoglycate

Steroid nasal sprays

Steroid nasal sprays can help to reduce sneezing and itching in the nose caused by hay fever. They may also help to reduce eye symptoms. Steroid nasal sprays take time to work, so you should start using them 1 to 2 weeks before you think your hay fever symptoms will begin. You will then need to use the spray every day over the hay fever period.

Commonly used steroid nasal sprays that are safe for those with acute porphyria include:

- beclomethasone
- budesonide
- fluticasone
- mometasone
- triamcinolone

These lists are not exhaustive so make sure you speak to your health care provider for more information about hay fever and what you can do to relieve the symptoms.

For more information, visit www.wmic.wales.nhs.uk/specialist-services/drugs-in-porphyria/ or call us on 02921 842 251.

Are you still receiving your annual SAFE drugs list? If not, please visit www.wmic.wales.nhs.uk/opt-in/

IDLE MUSINGS ON SUNSCREENS

As I sit and write it's the first Saturday of British Summer Time and the rain is incessant and even my new puppy refuses to go out!

But as a person with EPP I know that tomorrow the sky will clear and the sun will be out in all its "glory" and the battle will begin to try and get jobs done in the garden and normal day-to-day tasks which require venturing out into the outside world whilst all the time working out how not to get burnt by the "glorious" sunshine...

So, whilst reviewing my ever-growing collection of long-sleeved coats (with sleeves long enough to enable me to keep my hands covered), gloves (gardening and driving), solitary hat (nothing has worked quite as well as my Australian style bush hat – no corks, but does slip down over my eyes if it's warm!), my thoughts turned to the progress that has been made with sun screen....and whether there is now any alternate to the ineffective (for me) factor 50+ of a few years ago...but where to start my search for info...

Before I ask Dr Google

with its "guaranteed" factual accuracy, I start with the *BPA Skin Safety* page where I find a straight forward explanation as to why my previous "slap it all over...and repeat" factor 50 didn't actually work – it appears that that only worked against ultraviolet light and that "...reflectant sunscreens that are based on titanium dioxide or zinc oxide will be more effective."

The next question is what types of reflectant sunscreen are there? And will I look like a Test Cricketer with a white nose, lips and forehead? And although I know "it doesn't matter what other people think of how you look", it still takes a huge bucket load of self-confidence to go to Sainsburys looking like you have forgotten to wash off your Halloween face paint (in July...). So back to the *BPA Skin Safety page*... and yes there is a load of info on Dundee Cream and what I need to look for in other mineral sunscreens as well as a few alternate suggestions.

After a bit of search engine action and reading some customer reviews it appears that some products leave a



white residue, others absorb fully and some are tinted to reflect skin tone. But as stated by the BPA, and highlighted in some of the reviewers who state they ended up with "a nice tan", **even these sunscreens don't offer full protection**, and as those of us who live with EPP know, as well as us being very special people (I know this to be true as it was said once on an episode of the BBC's *Casualty*...), our tolerance of how much sun exposure we can take before we scurry off to the shady side of the road or scuttle back indoors is different for each of us.

So, will I take the plunge and try a sunscreen? I'm not quite sure, but with a new puppy to walk I am going to have to consider all options. Some alternatives available are shown below - top two have been shown to be at least as

effective as Dundee Cream and in some colours, even more so.

- Anthelios Mineral One (light, medium, tan, brown and dark brown), La Roche-Posay
- Anthelios Pigment Correct, La Roche-Posay
- Surf Mud natural zinc
- Invisible Zinc Broad Spectrum 50 SPF
- Neutrogena Sheer Zinc-Oxide Face dry-touch sunscreen 50 SPF

But whilst I ponder, the torrential rain has abated. So I will pull on my wellies and big coat and glory in a long dog walk in the glorious British Summer Time drizzle, where I don't have to worry about UVA and UVB rays, sun protection factors and that dreaded tingle that foretells that I have been out just a little bit too long...

by Claire Jarvis

¹ Ewan Eadie, Martin Josso, Rita Touti, Pascale Renoux, Robert S Dawe, Sally H Ibbotson, Commercial visible-light protecting sunscreens for photosensitive individuals, *British Journal of Dermatology*, Volume 188, Issue 3, March 2023, Pages 445–447, <https://doi.org/10.1093/bjd/ljac112>

THE BPA IS GROWING WELCOME GEORGIA AND CLAIRE

We are delighted to introduce two new members to the BPA Team. Firstly, welcome to the BPA's new freelance administrator, Georgia Newman. Georgia has been recruited to work alongside Liz and Sue for around 20 hours a month. Secondly, we are delighted to also have taken on a new administrative volunteer, Claire Jarvis. Claire is volunteering for a period of 12 months, whilst on a sabbatical from her other job and will be dedicating a similar number of hours to the BPA.

We've only been working with Claire and Georgia for a matter of weeks, but we're already seeing the impact that they are having on the work we are doing within the BPA. Please join us in welcoming them into the porphyria community.



GEORGIA NEWMAN



CLAIRE JARVIS

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

The BPA Newsletter is published by the BPA twice yearly.

Chairman: John Chamberlayne

Acting Treasurer: Anne Newton

Secretary: Richard Bennett

Strategic Lead of Operations and Advocacy: Liz Gill

Strategic Lead of Engagement and Advocacy: Sue Burrell

Patrons: Prof George Elder, Prof Mike Badminton,
Prof Felicity Stewart, Dr Penny Stein, Prof David Rees,
Dr Bob Sarkany, Dr Vicky McGuire

Viewpoints 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.

THANK YOU

A large portion of the BPA's income comes from fundraising and donations – we are always so grateful for the time taken and thought given by people who choose to raise funds or donate to the BPA. Your valuable support helps to enable us to do the work as we do.

We would like to take this opportunity to say THANK YOU to the Masonic Charitable Foundation for the kind donation of £100. We also received some very generous birthday fundraiser donations for Orla, via the Paypal Giving Fund, to the value of over £660, sending a huge THANK YOU to Orla and all of her friends who kindly donated!

We are sad to say that Amazon Smile is no longer functioning for charities to receive funds. But, we are very grateful to all of you who made use of it up to February this year, between November and January, the BPA received just over £60 – THANK YOU to all you savvy shoppers!

The BPA's Christmas Fundraiser of cards and baubles raised around £300, in the form of online orders and from a couple of Christmas Fete stalls – THANK YOU to everyone who purchased items or who ran a stall. If you would like to run a stall in 2023, please get in touch.

Finally, the BPA would like to offer a sincere THANK YOU to the family and friends of Helen Lane. In memory of Ms Lane, your generous and kind donations online and via the funeral directors raised a staggering total of more than £1,600. THANK YOU for thinking of the BPA at this sad time.

THANK YOU TO DAN

Danil Lopukhov has sadly had to step back from his role as BPA Treasurer and Trustee with the BPA. We would like to thank Dan for the time and effort dedicated to the BPA over the last year – we are very grateful and all wish Dan the very best in his future endeavours.

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 want to Gift Aid my donation and any donations I make in the future or have made in the past 4 years to the British Porphyria Association (Registered Charity No. 1089609). I am a UK taxpayer and understand that if I pay less Income Tax and/or Capital Gains Tax than the amount of Gift Aid claimed on all my donations in that tax year it is my responsibility to pay any difference. (I will advise the BPA if my tax status, name or address changes.)

DATE SIGNATURE