Issue 39 / **June 2020**

THE
BRITISH PORPHYRIA ASSOCIATION
NEWSLETTER

Registered Charity No. 1089609

LOOKING AFTER YOUR MENTAL HEALTH

While many of the population are coming to terms with a new way of living due to Covid-19 and learning how to cope with feeling isolated, this is not something new for the rare disease community, and even more so for patients with porphyria. People with all porphyrias, but especially EPP/XLEPP and CEP are likely to have suffered from aspects of feeling isolated before, and some will be amazingly skilled at adapting and doing things differently to try maintain some elements of normal day-to-day life.

Despite this, living with a rare disorder or coping with circumstances beyond your control can have a detrimental effect on mental well-being. Anxiety, stress, depression, low mood and emotional exhaustion have all been identified as occurring more in people living with long-term health conditions, along with an impact on quality of life. At the moment, there are additional impacts on mental well-being caused by Covid-19.

- **Stress:** Social isolation, reduction in physical activity, unpredictability in routine or work/school patterns can all contribute to increasing stress.

- **Anxiety:** Anxieties may arise over how to cope with changes to daily routines, how to get shopping, whether you can manage financially, ongoing care arrangements with health providers, or how to get support with medication/treatments.

- **Frustration, anger, heightened emotions or depression:** Shielding and social distancing can be frustrating. Restrictions on seeing family, friends, boyfriends/girlfriends and work colleagues can all have a negative impact on mood and feelings, and can also have an impact on sleeping patterns.

LOOKING AFTER YOUR MENTAL HEALTH (CONT.)

So, whatever the circumstances leading to a negative effect on mental well-being, it is important to acknowledge the subject and do all that we can to keep as happy and well as possible. It can be really easy to fall into unhealthy patterns of behaviour which in turn can make you feel worse. If you're feeling a little overwhelmed, why not try the following tips:

Focus on the things you can control

When you have little control over the big stuff, try not to spend time focusing on them. Try to divert your energies towards things you can control: what you eat, how much sleep or exercise you get, what you do with your day, and small and manageable changes you can take to make yourself as well as possible.

Try making a list of the things you want to achieve that day, month or year. Think about how you might be able to do them, then break the list down into manageable tasks or actions.

Connect to people, talk to others, ask friends/family for support

Think about how you can stay in touch with friends and family while at home. Phone calls, video calls or social media are all readily available; many of you will already be active on social media. Make use of them to connect with people you normally see often or even connect with old friends.

Look after your body

There's a known connection between physical health and mental well-being. Even if you are restricted to what you can do, try to make an effort to do some level of activity every day. For some people that might be a run or a cycle, but for some it will be simply climbing the stairs a few more times in the day.

If your diet needs attention, try to make small changes that you will be able to stick to. You can keep adding small changes once you know that you can manage the first ones. Keep it simple, try swapping a bag of crisps for a portion of fruit or exchanging white bread for brown!

Good quality sleep makes a big difference to how we feel mentally and physically, so it is important to get enough. Try to maintain regular sleeping patterns, avoid screens before bed and cut back on caffeine.

Be kind to yourself

When you've had a day when you've not managed to do all the things you wanted to do, or when you feel like you're not succeeding, try not to admonish yourself for it. Don't feel guilty and don't criticise yourself. Realise that it didn't work that day and try again the next day. Be kind to yourself!

Do something you enjoy

No matter what other demands are upon you, try to take a little time to do something that you enjoy. Whether that's taking a bath, reading, watching films, yoga, crafting or doing something more energetic, it's important to take some time for you.

Access support from other sources

- **Mind:** www.mind.org.uk
- **Anxiety UK:** www.anxietyuk.org.uk
- **NHS:** Every Mind Matters: www.nhs.uk/oneyou/every-mind-matters provides simple tips and advice to start taking better care of your mental health
- **No Panic:** www.nopanic.org.uk
- **SANE:** www.sane.org.uk/support
- **Young Minds:** www.youngminds.org.uk
- **Samaritans:** www.samaritans.org

A number of discussions at our festival event in Manchester focused on the subject of mental health and well-being. Presentations are gradually being uploaded to our YouTube channel, so please do keep checking to see what we've been uploading. https://tinyurl.com/ybhzf6ef.

EUROPEAN PORPHYRIA NETWORK (EPNET)

Epnet is a clinician-led, non-profit association which aims to improve healthcare services for porphyria patients. After a period of inactivity, Epnet reformed with new statutes and a new Board in October 2018.

One of Epnet's first tasks was to establish five new working groups:

- **Clinical: acute hepatic porphyrias**
- **Clinical: cutaneous porphyrias**

+ Both clinical groups aim to develop guidelines for the diagnosis, treatment and monitoring of the porphyrias. The aim is to publish recommendations that provide practical, unbiased guidance for clinicians and patients all over the world.

- **Laboratory:** aims to ensure consistency and quality in lab diagnosis, testing and reporting.
- **Research and development:** hoping to look into the penetrance of acute porphyrias and why some people with a faulty gene are more affected than others.
- **Website:** tasked with bringing the website back up to date and giving it a new fresh feel.

Well-known porphyria experts from the UK are involved in Epnet. Dr Mike Badminton (Cardiff) is on the Epnet Board and is on both clinical working groups, while Dr Penny Stein leads the acute porphyrias working group. BPA committee members are involved too. Liz Gill is the international patient representative on the Board and is a member of the acute group, while Antony Fearn is involved with the website working group.

In conjunction with the host country's porphyria specialists, Epnet also coordinates the International Congress on Porphyrins and Porphyrias (ICPP) which is a global event held every two years. For more information on the role of Epnet, please see: porphyria.eu.

THANK YOU

Porphyria Awareness Week 2020 was a massive success! The event was truly international with great engagement across many countries. Both the American and Australian patient organisations reached out to say how much they enjoyed our campaigns which flooded their social media channels. The BPA would like to say a massive and sincere THANK YOU to everyone who took part.

Thank you for all of the personal and enlightening stories that were shared – they really helped to put the spotlight on raising awareness of the true impact of living with porphyria by focusing on the challenges faced, as well as the determination shown by you all!

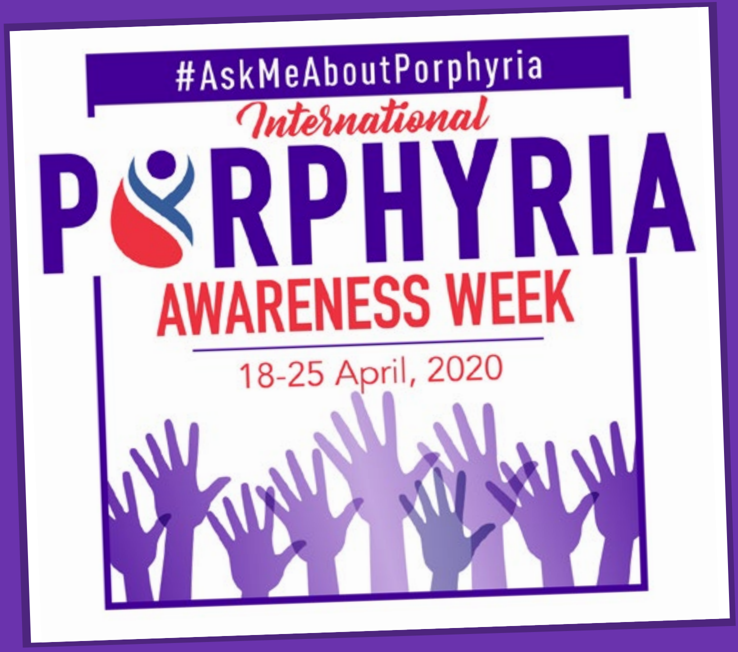
We'd also like to say thank you to all of those who have donated or been fundraising for the BPA. Despite the restrictions and strange times that we are all experiencing, and sporting events being cancelled or postponed, we have been truly astonished by the ingenuity of our fundraisers and level of engagement from our members, their families, friends and the wider community.

Whether you went red or purple, shared your story, created a video or got involved in the bRED activities, we want to say a huge THANK YOU for getting involved in an absolutely amazing porphyria awareness week!

- The Hutt family: David, Thirusha, Ettienne and Alexander ran and cycled the equivalent of 4 marathons (168 km) during Porphyria Awareness Week, raising an enormous £1079. Thank you!

- Thanks also to Paul and Frankie Worthington, who ran 120km during the 7 days of PAW, smashing their initial target to raise a fantastic £504.

- Bethan Rees took PAW to heart and made an outstanding contribution to the bRED events. To name just a few of the highlights, Bethan created a highly entertaining video while taking part in PE with Joe, hosted a quiz, did an evening of Just Dance, held a creative writing session, created a YouTube video on porphyria, oh, and dyed her hair a vibrant red! Thank you Bethan for raising nearly £550, a fantastic amount.



- The bRED4Porphyria Porphyria Awareness Week campaign via social media also raised over £170 – thank you to everyone who took part and made donations to the BPA.

- Evie and Ophelia Baldwin took the opportunity of no school to dye their hair purple to tie in with the international PAW colours. They absolutely crushed their initial target of £250, raising an enormous £1627! Wow, thank you so much! The purple hair looks amazing too!

- £638 in donations were also received in memory of Frank Murrill. £322 was also raised in memory of Susan Angela Rees and £135 in memory of Kath Walton. Thank you to all their friends and family for thinking of the BPA at this time.

- Finally, thank you to all who have nominated the BPA on Facebook for birthday donations. You have raised a wonderful £718 so far this year.

If we haven't named you individually here, it isn't because we don't care, we simply have limited space. Please be assured that all of your efforts and contributions, no matter how small or large, are massively valued and enable us to continue doing the work that we do. **THANK YOU TO YOU ALL.**

NEW Fundraising, Awareness and Social Media Working Group
Finally, we'd like to draw your attention to the new Working Group that developed this wonderful Porphyria Awareness Week plan, specifically: Victoria Harrold, Alicia Cawthorne and Natassja Chadwick who have recently become more involved in the BPA, without YOU this would not have been possible! Anne Newton and Karen Harris, two of the BPA's Trustees, also worked on the project alongside Sue Burrell. There were lots of Skype meetings, WhatsApps and emails and the whole BPA are extremely grateful for their dedication over the last few weeks.

The BPA would like to say specific and massive THANK YOUs to:

Victoria, for the countless posts managed on Facebook – your Facebook knowledge is so enlightening too. Thank you also for sharing your EPP story, your commitment to supporting others with EPP is amazing!

Alicia, for being our Instagram correspondent and for your commitment to telling your story and raising awareness. Alicia's story will be in Take a Break in a few weeks' time – it was also shared on *KentOnline* in April. What a fantastic effort Alicia – thank you.

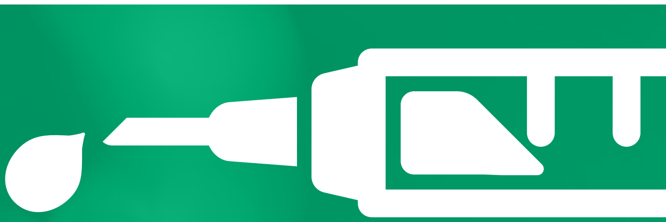
Natassja, thank you for being a total fundraising warrior, your ideas are amazing and your commitment to raising awareness is phenomenal (and the wonderful awareness video that you created on being a parent of children with EPP was so heartfelt and true). Thank you also for the continual fundraising efforts for the BPA – it is overwhelming, please keep up the amazing work! These three wonderful ladies have had such a positive impact in a really short space of time and we cannot wait to see what is planned next!

#THANKYOUNHS

Patients with porphyria rely on the NHS a lot, but the risks you're taking to care for us at this moment has us in awe of you all!

And for those patients with porphyria who are NHS staff or key workers, we say an extra special thank you to you too.

NEW DRUG DEVELOPMENTS



Givlaari
Following our Autumn update on the successful FDA marketing approval of Givlaari (Givosiran) for patients with acute hepatic porphyria in the United States, March 2020 saw Givlaari also being granted marketing authorisation by the European Commission (EC).

This is just the first step towards UK patients being able to access the preventative siRNA treatment. Approval means the drug is deemed safe and effective in the treatment of a disease. It is then down to national funding bodies to decide whether the drug is deemed cost effective enough for treatment to be funded. Givlaari will soon start going through the NICE (National Institute for Care and Excellence) process to try to obtain approval for use/reimbursement by the NHS in England. Similar procedures will follow for Scotland and Wales. *Learn more at* www.alnylam.com

Scenesse (Afamelanotide)
The NICE process for Scenesse has stagnated with no significant developments since our last newsletter, but we have been working hard behind the scenes to try to get things moving again. We have no news to announce at the moment, but will keep our communications going with NICE and with Clinuvel to make sure that EPP patients have a voice going forwards. *For more information, please see* www.clinuvel.com

Mitsubishi
Mitsubishi Tanabe Pharma Corporation is a company that we are closely watching. They are starting Phase III of a global trial into Dersimelagon (MT-7117) for EPP. This is a synthetic, orally-administered molecule in tablet form, which targets the melanocortin-1 receptor (MC1R) – the same receptor targeted by Scenesse (Afamelanotide). It is a new type of medicine which we hope might reduce phototoxicity in patients with EPP/XLEPP. The company has adopted a different way of determining efficacy, which is the average daily time (in minutes) to first prodromal symptoms associated with sunlight exposure. Prodromal symptoms are the ones that you get before the main problem develops. With EPP they are often a tingling warning that you need to avoid further light exposure.

This trial has already started in the United States, and we are hopeful that it will soon expand to the UK and Europe. We'll keep you posted as we hear more.

VACANCY:
VOLUNTEER TREASURER/ TRUSTEE

We'd like to share news of an exciting opportunity. We are looking for a Treasurer to join our Board of Trustees following an extended handover period. 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. Find the full details at www.porphyria.org.uk/vacancies

Applications should be made via John Chamberlayne, the BPA Chairman chair@porphyria.org.uk. Please provide your CV, along with a covering letter stating why you wish to work for the organisation, how your skills would add value to the board and any other relevant information.

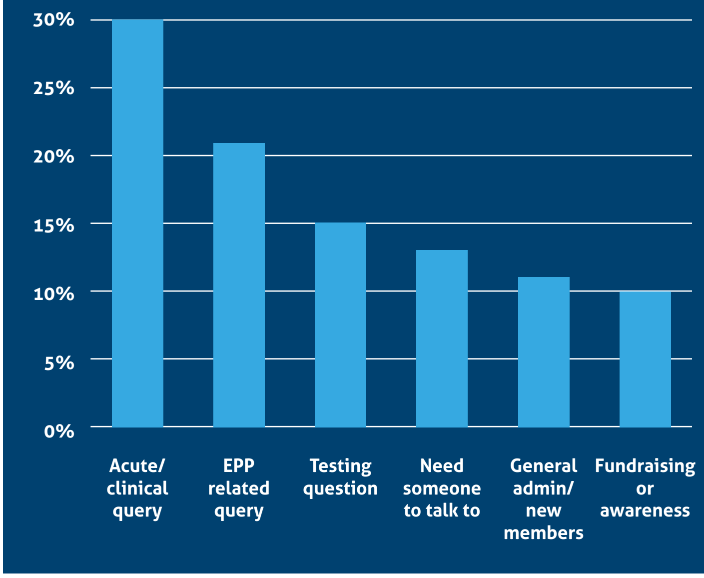
HELPLINE USE

We know that our members value our email and telephone helplines. But we thought it would be interesting for members to know what types of queries we regularly answer. We monitored the types of calls/emails we got over a six-month period and categorised them according to the chart below.

We also help to answer patient questions via social media, but these queries haven't been included in this analysis.

We're always keen to hear feedback, so if you have any comments or suggestions on how you currently receive advice, or how you might like to raise questions and seek advice, do let us know.

Helpline Query Topics



EVENTS 2020

Following the amazing success of the festival, we had hoped to bring you a great event in September 2020 in Milton Keynes. However, with all of the uncertainty around Covid-19 and the fact that many of our members fall into vulnerable categories, we feel it would be unwise to pursue a face-to-face gathering this year. So, with that in mind, we are looking to hold a completely different type of occasion. We are hoping to put together a virtual event that all will be able to attend, no matter what the restrictions on movement. We haven't got any concrete plans yet, but we are hoping it will be on 19 September 2020. Keep a look out for communications in due course.

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.

A Film about EPP

Andrea Minoglio presented an amazingly moving documentary: ‘Overexposed: a film about light and erythropoietic protoporphyria’ at the International Congress on Porphyrins and Porphyria (ICPP) in Milan last September: *https://vimeo.com/359771483*

The film has been selected for two film festivals. If you would like to vote, you can do this here: www.unosguardoraro.tv/festival/edizione-2020/

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
☐ Patient events/information
☐ Newsletters
☐ 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