



Q&A: Skin porphyrias interactive session

We'd like to thank all attendees for posing so many great questions and getting fully immersed in the day.

Many questions were answered during the event – but any that we didn't get time to answer or questions that we thought were worth reiterating the responses to are included below.

Skin porphyrias

EPP and PCT THERAPIES



<https://youtu.be/7Cxb5m41xeo>

Has Scenesse been proved to be safe to use? Yes, Scenesse has been proven safe to use and approved as so by the European Medicines Agency and Federal Drugs Agency in the USA.

Any chance of Scenesse being approved in the UK? NICE made a recommendation not to approve Scenesse some time ago. We still hope that one day it will be, but at the moment, there is effectively a stalemate, largely regarding costs, between the manufacturing company, Clinuvel, and NICE where communications have come to a halt. We have worked in the background to get the process moving again, but so far efforts from the BPA and international representatives have not been successful.

Which other medicine treatments than Mitsibishu are being tested? At the moment, there isn't anything else close to being trialled, but research is always ongoing for the next thing.

What is the skin cancer risk for UV therapy? There has been a study published recently in the British Journal of Dermatology that has shown no additional risk of skin cancer with phototherapy treatment in a follow up of 20 years.

MONITORING (ROUTINE TESTING), VITAMIN D AND IRON

EPP: What tests?

MONITORING & VITAMIN D



<https://youtu.be/E6Kz93kSVZQ>

I may have liver complications, or another liver condition. Is there a liver function blood test? I've had fibroscans already which show damage. Liver function performance can be done from a blood test and if needed other tests such as scans will be used to gain more detail about any problems.

Thinking about guarding against osteoporosis and thinner bones in later life.... What types of activities would be most advantageous to the EPP patient? Sports and activities that are largely aerobic such as swimming, or those that involve more power and strength related exercise such as gymnastics or climbing. Impact/load/weight bearing exercise such as walking, running, cycling, dancing, weights, climbing and weightlifting are the best for strengthening bones, rather than swimming.

We are considering not travelling to London this year for my daughter's appointment at Guys because of coronavirus. Can she have bloods taken at our GP this year if they are stored correctly? If you or the GP are able to send the (light protected) blood samples to the lab this should be possible. Communication with Guys and the GP will be important to enable this to happen.

Do very high levels of free protoporphyrin levels affect sensitivity to the sun and what do they mean?

There is some evidence to suggest that the people with higher levels (25-50mmol), may have more severe symptoms that happen faster and last for longer, whereas those with lower levels, e.g. 5-15 or 20mmol may have milder symptoms and take a longer time to react as well as less time to recover. In research mentioned by Dr Sarkany, there was a difference between the two groups, but the differences weren't straight forward and this might just be one factor in amongst factors such as differences in the body and where the blood vessels lie, as well as pain thresholds too.

Vitamin D is also stated to be important for the immune system, does this put EPP patients more at risk of illness? As with other vitamins, it is important to have sufficient vitamin D to maintain a healthy body. Any deficiency can put people more at risk of illness, so it is important that levels are adequate. <https://www.nhs.uk/news/lifestyle-and-exercise/vitamin-d-immune-system-boost/>

Can you remind me how much Vitamin D I need, I've just checked my multivitamins and it says vitamin d is 10ug, is that enough or do I need a separate tablet? UK specialists routinely put photosensitive patients on a standard dose of 1000U or 25ug/day vitamin D3 long term, unless their results are fine, which is rare. Please check with your GP or specialist to get advice specific to you and your results.

What causes liver damage in EPP? Can more sun exposure cause liver damage? The high levels of protoporphyrin irritates the liver and in particular causes damage to the bile ducts. More sun exposure does not cause liver damage. Experiments have shown that protoporphyrin damages the liver even without light.

Is it safe for patients with EPP to have an iron-rich diet? Dr Sarkany recommended liaising with a dietician and porphyria specialist on an individual patient basis to see whether any particular iron-rich diet comes anywhere near the levels that would be seen in iron supplementation as if it does, it would be considered with the same caution as iron supplementation. Iron is treated with caution in EPP because the effect is unpredictable in EPP, where some people get more skin sensitivity and also problems in the liver, so need to be monitored.

Would you suggest sleeping pills for EPP pain? There's no reason why an EPP patient should or shouldn't use sleeping pills, just like for anyone with any health condition causing pain. A discussion ensued over patient preferences for this method and some people have found it to be successful as it let them sleep while recovering. It was also acknowledged that when sleep eventually is possible after a reaction that it is a deep, heavy sleep and that the sleep does seem to aid recovery.

DAY-TO-DAY MANAGEMENT

EPP: what are the benefits of trying to

EXPOSE THE SKIN TO MORE LIGHT?



<https://youtu.be/rlwaE0AYnnY>

Question for Antony: Nice tan, mate! Where did that come from? Also, Dr Sarkany mentioned that some people seem to have a greater tolerance for light exposure than others - hours, rather than minutes. Are two distinct 'levels' of EPP recognised by the medical community, or is a continuum?

Antony explained his gradual exposure philosophy of short but regular exposure to some sunlight, which he has found can work to toughen the skin slightly and provide a tan to protect – although he also reported being caught out and experiencing painful episodes when he pushed the boundaries too far, especially in the spring and early summer, whereas later in the year, he felt it offered some level of protection.

An interesting observation was noted by Kirstine Belongie from Mitsubishi Tanabe that people on placebo who thought they were on the drug went out 20mins more than the placebo group who believed they were on the placebo. This group managed to change their behaviour by about 20 minutes without pain which led some of these patients being able to build some resistance in the skin.

This also led to discussions about carefully planning every day around where the sun might be, what the detailed weather forecasts predict and what protection you can find during the day, such as woodland or buildings.

A patient also shared an app for your phone *EVO Lightspectrum* that can help patients to analyse light exactly at that moment. He felt it had helped him a lot this year.

There were numerous questions relating to EPP reactions from computer screens, TVs and fluorescent lighting, which a couple of people told us caused lots of problems for them.

Dr Sarkany noted that the measurable intensities of visible light from internal electrically powered devices is a lot lower than external sunlight, and it is quite rare to see internal light causing EPP problems. However, Dr Sarkany did note that some people with extraordinarily high levels of protoporphyrins, such as from liver complications, did experience problems with internal light, and so if patients are experiencing this all the time, then it might be advisable to check back with a porphyria specialist to see whether anything else could be at play.

It was also acknowledged that patients can be affected by this type of light when already suffering a reaction as the skin is hypersensitive at this time.

If your EPP is sufficiently severe to be affected by the light from your phone or computer screen, it is possible to reduce the amount of blue light that your device gives off. Some phones have a setting to do that in settings / display called blue light filter, or twilight on Android, night shift on iPhones, or night light on Windows phones. For recent PCs, night shift or night light is available in display settings. If your phone or PC doesn't have that, then you can download a suitable app – search for blue light filter in your app store (play store, app store or Windows marketplace) or Google. All of these have been designed to help with getting to sleep, but should be just as useful if you need to reduce the blue light emitted to help reduce symptoms of EPP.

I note suncreams are used by a lot of patients, are there any suggestions for a good one to use? I never have any luck. I tried Dundee Cream but struggled with this. Sun creams seem to offer limited if any benefit to EPP, especially generic off-the-shelf sunscreen. They don't fully block the wavelengths of light that trigger EPP. Creams that contain some form of physical barrier such as zinc oxide and especially Dundee Cream offer some level of protection. <http://www.porphyrria.org.uk/skin-safety/>

I guess the reason patients use them is that even for patients like myself who do build up some resistance our sensitivity to light is so great, and the consequence of a very painful reaction so high, that we will use anything at our disposal. even if it only makes a 1 or 2% difference. Without an effective treatment in the UK every such gain is like gold dust (Antony).

Our members have also 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 Sarkany also noted the following creams that he has come across recently that claim to contain iron oxide and may be useful for blue and violet light sensitivities – these are as yet untried, but may be worth experimenting with – 360 Heliocare Sunscreen Gel; 360 Heliocare Sunscreen Gel Oil-Free 50; and Murad environmental shield city skin.

Can EPP make you more sensitive to even mild temperature changes? Especially in a flare up. EPP patients report becoming acutely sensitive to slight changes in temperature during flare ups. however rather than temperature, it is likely that even slight changes in levels of light that trigger EPP are what we are sensitized to.

What about operating theatre lights, I have read that they can be an issue? The key is making the team aware of potential problems, especially in lengthy operations, where a light filter has been used in operations such as liver transplants in the past. For emergencies it may be worth carrying a small info card... something we could look into developing for patients.

Can you tell us a little about

GENETICS AND FAMILY PLANNING IN EPP?



<https://youtu.be/MkbWln9i2Jg>



<https://youtu.be/TGv1mb5T9w8>



<https://youtu.be/4lwh45ESmTw>

Natassja, what has been your experience when explaining your children's condition to their school/teachers? Were they understanding and able to help cater to their needs?

My first tip would be to show someone you want to talk to about EPP and your children is a video of a reaction. I use this one:

<https://www.youtube.com/watch?v=KYyOai2R7q8&list=PLtdPRzDKPvCjzR72qJI7yemWa5SvRaa&index=28>

I make sure I meet with new teachers and the school every year and talk them through what happens when someone with EPP is exposed. Look at the worst case scenario of an attack and the pain it causes. I also have a letter from their specialist which explains the need for school to cooperate and be understanding with their condition. I outline what needs to be done to keep them safe, we have a care plan which is on the wall in their classrooms for supply teachers to also be aware of. I outline what I expect them to do to keep my children safe. It is really important to build and maintain your relationship with the school, be assertive but also be understanding of restraints and work together to overcome any obstacles.

I have had issues and I haven't always been happy with my children's care, but It's important to see things from the school's perspective too. Try work together but if you think there are certain things that can be improved on, don't be afraid to push for change. (Natassja)



https://youtu.be/luF_HSSuwAs

Have you any tips on dealing with feelings of rage and anger or feelings of wanting to hide away? Dr Rukshana Ali encouraged people to start noticing what they are feeling on a day-to-day basis, as usually the volcano of anger erupts after a while of building up. It's about learning to recognise those feelings of irritation, tension or being overwhelmed, then thinking about what you can do to improve that. She suggested the 5-4-3-2-1 technique. So, start thinking about five things you can see, four things you can touch, three things you can hear, two things you can smell and one thing you can taste. This can help you to ground yourself and think about what is causing the problem. Natassja also suggested taking time out to walk away, meditation music or calming music to take time to reduce the immediate anger and create calm, so that it's possible to process those feelings.

How bad can an EPP attack get? Can it be life threatening? Not that we are aware of. Although it was noted that infections can arise if there has been serious skin damage from exposure – so to take care with looking after any wounds.



<https://youtu.be/FBOiB7rHxgY>

Are there any studies or research on CBD lotion for EPP pain? It takes my pain from a 10 to a 5 in seconds. After 32 years it's the only thing I've found that works significantly during a reaction. An interesting discussion ensued on this topic. Deepak had a patient who tried CBD oil in the hope of reducing fatigue and some level of pain. UK NICE guidelines state that the quality of evidence on CBD oil as a pain relief option isn't sufficient for them to recommend as a treatment. Currently it is considered a health food and manufacturers are not compelled to do any scientific studies or take any particular action regulations-wise, other than to show that it contains less than 0.1% THC – i.e. it is not psychoactive. Therefore, there are no good studies on the topic. If people are wanting to try it as an option, Deepak recommended obtaining from a reputable supplier such as Holland and Barrett – starting with the lowest possible dose in a tablet or under the tongue form. Sticking to one brand. Gradually climb the dose after a month and if no benefit, stop at that point.

Katherine also noted that she found aromatherapy cream containing lavender and peppermint which gave some pain relief when skin is sore.

Dr Sarkany reported that experiments have shown that anxiety and stress levels can lengthen the time an injury takes to heal on the surface of the skin and it might be that this is also the case with the blood vessels.



<https://youtu.be/IQ39kNmRHzM>

Do people with EPP get more sensitive as they get older? I find my daughter is more sensitive this year. An ongoing study in the Danish population appears to be showing that protoporphyrin levels do increase as they get older and during the teenage years, and so this may correlate with children getting more sensitive as they grow up, but it should even off when they get to adulthood.

Holidays: What do you think about going on holiday to countries such as Spain? My 14-year-old daughter with EPP has never been abroad yet and sadly missed out on a school trip because of it. Do any families manage this? In the UK, patients on the meeting reported using venues such as Center Parcs that can offer more protection from the light with wooded areas and also lots of indoor activities. Cruise ships were also noted to offer a range of activities indoors and outdoors, where patients and their families can all do activities separately if needed. Antony recalled extremely negative experiences with the Mediterranean and Spain, especially beach-type holidays. But has found some active-type holidays where you may be able to protect yourself more with suitable clothing etc. to be more successful. He also visited Malaysia and Borneo, where there was lots of rain, cloud, and heat and lots of vegetation which also offers protection. Also these countries tend to wear more clothing and hats, so an EPP patient tends to fit in more and feel less awkward. The overall feeling was that any travel must

be done with the EPP patient in control and able to feel that there is somewhere to seek shade and keep safe.

PCT, VP and HCP question

Many patients suffer from severe blistering, what is the best way to look after these to reduce infection? Especially when they can be the size of a 50p piece? Keep it clean by running clean saline or clean cool water on it, cover with a simple gauze or other non-adhesive dressing. If it is weeping pus or more painful and red than usual it may have become significantly infected and you will need to see a doctor.

Also some patients have noted that the following cream may be useful at soothing skin:

[https://www.boots.com/eucerin-aquaphor-soothing-skin-balm-40ml-10131555?cm_mmc=bmm-buk-google-ppc-_-PLAs-_\(GB%3AWhoop!\)%20Eucerin-_\(GB%3AWhoop!\)%20Boots%20Shopping%20-%20Category%20-%20Beauty%20-%20Mobile&gclid=Cj0KCQjw_r3nBRDxARIsAJIjleGj5qSY3oUviHFyOPix2zv7oU--e0w1RTtFugnOd9Bad2y2znMMCQQaAjXyEALw_wcB&fbclid=IwAR3vNWJBca8pffxJ0vUZ0CB_M8N-f2N5DSy9apthHKamyGTvrVvyotk7z5o](https://www.boots.com/eucerin-aquaphor-soothing-skin-balm-40ml-10131555?cm_mmc=bmm-buk-google-ppc-_-PLAs-_(GB%3AWhoop!)%20Eucerin-_(GB%3AWhoop!)%20Boots%20Shopping%20-%20Category%20-%20Beauty%20-%20Mobile&gclid=Cj0KCQjw_r3nBRDxARIsAJIjleGj5qSY3oUviHFyOPix2zv7oU--e0w1RTtFugnOd9Bad2y2znMMCQQaAjXyEALw_wcB&fbclid=IwAR3vNWJBca8pffxJ0vUZ0CB_M8N-f2N5DSy9apthHKamyGTvrVvyotk7z5o)