

# Skin Protection in Cutaneous Porphyria

*This leaflet is intended to compliment the information provided in the individual patient information leaflets published on the EPNET website. The information in this leaflet is based on best available evidence and the consensus of the cutaneous porphyria working group of EPNET, <https://porphyria.eu>.*

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## 1. Why do porphyria patients have skin problems (photosensitivity)?

Porphyrias resulting in skin problems are known as cutaneous porphyrias. The skin problems are caused by an increase in chemicals called porphyrins circulating in the bloodstream, which take up energy from visible light causing a phototoxic reaction in the skin. There are two main types of skin reactions in the cutaneous porphyrias; fragile skin with blisters and acute painful photosensitivity.

## 2. Will I have skin problems for the rest of my life?

In many cutaneous porphyrias it is not possible to reduce the circulating porphyrins associated with the skin problems. However, the severity of symptoms can vary greatly between individuals. Skin problems in some patients with variegate porphyria and hereditary coproporphyria can resolve spontaneously.

Effective treatments are available for porphyria cutanea tarda, which can normalise levels of circulating porphyrins resulting in resolution of the skin problems. In most patients symptoms will not return, unless there is a relapse in the condition which has caused the PCT (see PCT patient leaflet).

In both erythropoietic protoporphyrias and congenital erythropoietic porphyria skin symptoms are generally present throughout life.

There are measures that all cutaneous porphyria patients can take to reduce the symptoms caused by photosensitivity.

### 3. How can I reduce exposure to sunlight?

Patients with a porphyria causing fragile skin and blistering may not be aware that their symptoms are caused by light exposure. For porphyria patients with fragile skin and blistering, the damage occurs slowly and the link is not always obvious.

It is very different in erythropoietic protoporphyria (EPP), X-linked protoporphyria (XLP) and to a certain extent congenital erythropoietic porphyria (CEP), where the effects of light exposure can be apparent within a few minutes.

Practical measures include:

- Avoiding the brightest time of the day outdoors (11am-3pm). There may be occasions when bright artificial lights need to be avoided as well
- Seeking shade wherever possible
- Being aware that light reflects from different surfaces (e.g. sand, water, snow and concrete) and you may not be protected even under a parasol, as light bounces in many directions from these surfaces.
- Making sure desks (at school or work) are away from unshielded windows (see window films below).
- Physical barriers to light exposure as described below

### 4. Physical barriers to light exposure

- **Clothing** should have a weave that limits the passage of light (check by holding up to light and see how much shines through – less is better). Avoid thin, light coloured fabrics. Loose fitting long sleeved tops and long trousers in darker colours give good protection. Shirts with collars and scarves can help to protect your neck. Clothes give less protection if they are tight fitting and become wet or stretched.
- **Hats** should be broad brimmed and dark in colour to protect your nose, ears and neck. Baseball caps expose your neck and straw hats let too much light through.
- Avoid open toed **shoes** such as sandals or flip-flops.

- Consider wearing **gloves** when exposed to sunlight.
- Over the counter **sunscreens** protect the skin against ultraviolet light (sunburn caused by UVB and increased risk of skin cancer caused by UVA) and are not generally effective in cutaneous porphyria, as visible light triggers porphyrins in the skin. However some widely available sunscreens zinc oxide which is added to protect against blue light, which may provide some protection in porphyria.
- Dundee cream is a visible light blocker, which comes in 3 different colours (Beige, Coral and Coffee). The different shades can be blended to suit your skin type. Dundee cream is available on prescription from hospital pharmacies. It does not suit everyone in terms of the way it looks or feels.
- **Window films:** Only UVB is blocked by clear windows; UVA and visible light will penetrate clear glass. Specialist clear window film blocks UVA, but not visible light and may offer some protection (e.g. Dermagard, UVCL). The most effective film for visible light induced cutaneous porphyrias is amber tinted specialist window film (ARC UVTA, TA81 Madico). Suppliers may differ in other countries.
- **Car windows:** The regulations for car window screens should be checked in your own country. As an example, in the UK window films must transmit 75% of light through the windscreen and 70% through front side windows. There are no restrictions for rear car windows, but this should be checked in other countries
- Umbrellas with a dark or reflecting surface

## 5. How can I protect my skin further?

Fragile skin occurs in areas exposed to visible light; most commonly the backs of the hands, forearms, feet, face and scalp. Prevention through photoprotection with clothing and visible light blocking sunscreen is effective in some patients, as discussed above. The backs of the hands are particularly vulnerable to damage and gloves should be worn whenever you are in direct sunlight, and in the blistering porphyrias, when undertaking manual tasks. There are a variety of options available, from heavy duty work gloves (e.g. for gardening) to cotton gloves for lighter work. If blisters occur, particularly if they are large or painful, they may be pierced with a sterile needle and the roof left in place as a natural dressing. This decreases the likelihood of further skin damage and secondary infection. Hand washing will also reduce the risk of skin infections and a regular emollient ointment should be used after washing as a barrier to protect your skin.

## 6. Why is Vitamin D important?

Vitamin D is essential for healthy bones, teeth and muscles. Low vitamin D levels can result in illness (e.g. osteoporosis/osteomalacia in adults and rickets in children). Sunlight avoidance and photoprotection in cutaneous porphyrias can lead to vitamin D deficiency, as sufficient quantities are not being made by skin exposure to the sun. You should have your vitamin D level checked at least annually. Levels are lowest in the winter months, but cutaneous porphyria patients should take supplementation throughout the year. Depending on the vitamin D blood result and on how well the vitamin D increases to normal will determine what dose of vitamin D is to be taken as a supplement. Most patients find that they require long term supplements of 1000Units (25 microgrammes) of vitamin D3 daily. A bone mineral density measurement should be considered at least once, in all persons with low sunlight exposure.

## 7. What treatments are available for cutaneous porphyria?

**See advice in disease specific patient information on porphyria cutanea tarda (PCT), congenital erythropoietic porphyria (CEP), erythropoietic protoporphyria (EPP) and X-linked erythropoietic protoporphyria (XLP).**

## 8. What other action can I take?

Porphyrias are rare diseases, which may not be well understood by the wider public and other people may not be aware of how to offer support, particularly at school or in the workplace. Your porphyria specialist should be able to provide a letter to your employer or head teacher explaining your diagnosis and recommendations for measures to protect you from developing symptoms and allow you to achieve maximum participation at work or school. Patient associations can be a good point of contact for information, networking and support. Porphyria patient groups in different European countries are listed on the European Porphyria Network website, <https://porphyria.eu/content/patient-organizations> and the Global Porphyria Advocacy Coalition (GPAC) website, <https://www.gpac-porphyria.org>.

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