

hormone preparations are very dangerous and must be avoided. The Mirena coil is a regularly used contraception method for acute porphyria patients.

Pre-menstrual symptoms: Unlike in acute intermittent porphyria, premenstrual attacks and repeated acute attacks are unusual in HCP.

Pregnancy: Though nearly all pregnancies are uneventful, there is a small increased risk of having an acute attack during or after pregnancy. It is therefore very important for the doctors providing care during pregnancy to know that you have HCP.

How is an acute attack diagnosed?

In a patient who is known to have inherited HCP, it may be difficult to decide whether an illness is an attack of HCP or something else. In this situation, urinary PBG (porphobilinogen) can help your doctor to decide whether you are suffering from your porphyria or some other illness. It is important that people with porphyria do not assume that all of their illnesses are porphyria related, as common but potentially serious conditions, such as appendicitis, may be overlooked.

Treatment of acute attacks

Early recognition of an attack allows early treatment. The early symptoms are often easily recognised by those who have previously experienced an attack. An immediate increase in carbohydrate intake (sweet or starchy food and drinks) may help to reduce the severity of the symptoms.

If symptoms are severe, or don't settle within 24 hours, you should contact your doctor. This will allow for:

- Confirmation of the attack by measuring PBG.
- Early start of specific treatment, e.g. haem arginate.
- Treatment for the various symptoms such as drugs to relieve pain and nausea, or actions to maintain an adequate intake of calories.

Your doctor should contact NAPS (National Acute Porphyria Service: *see back page*) who will give advice about treatment and provide haem arginate if needed.

Useful contact details

BPA telephone helpline: 0300 30 200 30

BPA email helpline: helpline@porphyria.org.uk

European Porphyria Network: <https://porphyria.eu>

Rare Connect: www.rareconnect.org

British Association of Dermatologists: www.bad.org.uk

Medic Alert: www.medicalert.org.uk

UK Porphyria Medicines Information Service (UKPMIS)

UKPMIS provides advice and a list of SAFE drugs which can be downloaded at www.wmic.wales.nhs.uk/porphyria_info.php. They can also be contacted by telephone on 029 2074 3877 or 029 2074 2251.

National Acute Porphyria Service (NAPS)

NAPS provides clinical advice and haem arginate where appropriate for patients having either one-off acute attacks or recurrent attacks of porphyria. There are two full NAPS centres and two associate centres:

- King's College Hospital, London
- University Hospital of Wales, Cardiff
 - * Salford Royal Hospital, Salford
 - * St James University Hospital, Leeds

Initially, your doctor would need to contact the emergency number at the University Hospital of Wales: 029 2074 7747. This 24/7 number should be used at all times for new patients, and out of working hours for existing NAPS patients.

Specialist porphyria laboratories

- For an up-to-date list see the British and Irish Porphyria Network (BIPNET) website: www.bipnet.org.uk.

Protective clothing

A range of protective clothing and products can be found from:

- SunSibility: 0208 224 2299 or www.sunsibility.co.uk
- Rohan: 0800 840 1411 or www.rohan.co.uk
- Sun Togs: 01733 765030 or www.sun-togs.co.uk
- Equatorsun: 01932 230907 or www.equatorsun.com

Dundee Cream

Dundee Pharmaceuticals, Ninewells Hospital, Dundee. DD1 9SY. Tel: 01382 632052.

British Porphyria Association

Charity No: 1089609



Hereditary Coproporphyria (HCP)

Information for patients



Introduction: what is porphyria?

The 'porphyrias' are a group of rare disorders. Most are inherited and result from a faulty gene leading to difficulty making a chemical called haem. Haem is a constituent of many important proteins in the body. Haem precursor chemicals accumulate which can cause severe medical problems.

Hereditary coproporphyria (HCP) is one of four porphyrias that may present with 'acute attacks'. Attacks are uncommon and often difficult to diagnose. In addition to acute symptoms, the skin may also be affected in people with active HCP. People with HCP may find that areas of skin exposed to sunlight become fragile, leading to blistering, small tears or bleeding — skin should therefore be protected.

Occurrence

HCP is quite rare in the UK, although the exact occurrence is unknown. The prevalence of symptomatic HCP (i.e. those suffering from attacks) is thought to be one in a million people.

Most people with HCP will never have any problems due to their condition. Almost all of those that do will have acute attacks, and about 1 in 4 of those with an acute attack will also have skin problems. It is very rare for people with HCP to have skin sensitivity alone, but this is more likely in people who also have liver or gall bladder problems. **Everyone with HCP should avoid triggers for acute attacks.**

Skin problems

Porphyrins can build up in the skin particularly during acute attacks, causing sensitivity in areas exposed to sunlight. Exposed skin is most affected, particularly the backs of the hands, face, legs, the scalp and the tops of feet (due to wearing sandals). Small blisters can appear or small white dots (milia). Skin is more fragile than usual so scrapes and cuts take time to heal.

If you have skin sensitivity you should take the following precautions (not everyone with HCP will find this necessary, adjust your actions to suit you).

- **Avoid direct sunlight** as much as possible.
- **Protect hands** with gloves when bumps and scrapes are likely. Broken skin should be washed and protected with a dressing to prevent infection.
- **Protect exposed skin** from sunlight with suitable clothing (long sleeves, light cotton gloves, wide-brimmed hat). (*See back page for examples of UV protective clothing suppliers.*)
- **Try sunscreens.** Although ordinary sunscreens are not very effective, those with a high star rating, which is a measure of UVA protection, may provide some protection. Special opaque sun creams that block visible light may help if skin problems are severe, such as Dundee cream, which must be prescribed by your GP or porphyria specialist (*see back page*).

Acute attacks

An acute attack usually lasts for no longer than one or two weeks. However, if severe neurological complications such as motor paralysis occur, recovery will be gradual but slow.

Typical features of acute attacks:

- **Severe pain** in the abdomen, back, arms or legs.
- **Nausea, vomiting and constipation.**
- **Low sodium (salt) levels** in the blood.
- **Pulse rate and blood pressure may increase**, but rarely to dangerous levels.
- **Confusion** may occur during an acute attack.
- **Convulsions and muscular weakness** which may lead to paralysis can arise.

How can I reduce the risk of attacks?

Acute attacks are extremely rare before puberty and most people affected by attacks will only have one or a few attacks in their entire life. However, people with HCP need to take a few simple precautions to reduce the risk of attacks. Acute attacks can usually be reduced by avoiding triggers.

Typical acute attack triggers:

- Certain drugs, including prescribed, over-the-counter or herbal remedies
- Alcohol
- Fasting - including dieting and gastric infections
- Hormones
- Infections, viruses and stressful situations

Drugs: The most common trigger for acute attacks in HCP is drugs. The UK Porphyria Medicines Information Service (UKPMIS) has compiled a list of SAFE drugs for easy reference. The list is updated annually and is available to view online. Patients/doctors can call **029 2074 3877** (*more details overleaf*). Drugs not on the safe list should only be taken after expert advice from UKPMIS or a porphyria specialist.

The Norwegian Porphyria Centre (NAPOS) drug database www.drugs-porphyria.org contains further detailed information on prescribing in acute porphyria. NAPOS classifies drugs into five categories according to their potential to cause symptoms.

ALWAYS check the safety of any medicine with your doctor/pharmacist using the SAFE list

Even though acute attacks are very rare before puberty, it is safest if children also avoid all drugs that are not known to be safe in porphyria.

Alcohol is a common trigger, especially in high volume, so the best advice is don't drink or keep intake low.

Diet: Low calorie diets, or prolonged periods with little food, may provoke acute attacks. It is therefore important to keep to a normal diet with regular meals, eating enough to maintain a desirable body weight.

Particular issues for women

Women are at least three times more likely than men to experience an acute attack, mostly due to female hormones. If possible, women with HCP should avoid oral contraceptives and HRTs that contain progesterone or progestogens. Injectable and implantable long-term