

hormone preparations are very dangerous and must always 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 variegate porphyria.

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 VP.

How is an acute attack diagnosed?

In a patient who is known to have inherited VP, it may be difficult to decide whether an illness is an attack of VP 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/specialist-services/drugs-in-porphyria/. 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:

- Decathlon: www.decathlon.co.uk
- 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

Dundee Cream

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

British Porphyria Association

Charity No: 1089609



Variegate Porphyria (VP)

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.

Variegate porphyria (VP) is one of four porphyrias that may present with 'acute attacks'. Attacks are uncommon and often difficult to diagnose. In addition to acute symptoms, in VP the skin may also be affected. VP may cause areas of skin exposed to sunlight to become fragile or to blister — skin should therefore be protected. In VP, the skin changes and acute attacks may occur at different times.

Occurrence

VP is quite rare in the UK, but is quite common in South Africa.

Most people with VP will never have any problems. Around 3 in 5 of those that do will have skin sensitivity only, while 1 in 5 will have acute attacks only and the remaining 1 in 5 have both acute attacks and skin problems. **Everyone with VP should avoid triggers for acute attacks** (see third column for more detail).

Skin problems

Porphyrins can build up in the skin at certain times, causing sensitivity in areas exposed to sunlight, particularly the backs of the hands, face and legs, the scalp and the tops of feet (due to wearing sandals). Small blisters can appear. When they break, they leave narrow brown hollows (often called 'ice pick lesions'). They gradually fill up and disappear. Occasionally, VP can cause larger blisters that may be mistaken for porphyria cutanea tarda (a skin porphyria). VP can also cause small white dots (milia) which don't go away.

Skin fragility is common, and scrapes and cuts take time to heal. Wash wounds carefully and protect with a dressing to prevent infection. Hands can be protected with gloves when scrapes are likely. If you have skin problems, you should look after your skin carefully.

Avoid direct sunlight as much as possible, even sunlight that passes through window glass in your home or car.

Clothing: Protect your skin by wearing clothing with long sleeves, light cotton gloves and a wide-brimmed hat. Not everyone will find this necessary or acceptable; adjust your clothing as it suits you. Many companies now provide sun protective clothing (see back page for examples).

Sunscreens: Ordinary sunscreens are not very effective. The ones that help the most are the thick, opaque preparations with high protection factors (often containing titanium dioxide) that block visible light. Dundee cream is designed for skin porphyrias. Your GP needs to prescribe it for you. Your GP/pharmacist will need to contact Dundee Pharmaceuticals (see back page).

In the UK, the SPF (sun protection factor) tells you how effective the sunscreen is for UVB, and the star rating provides a measure of the UVA protection. High star rating sun creams provide limited protection.

Acute attacks

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 occur.

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.

How can I reduce the risk of attacks?

Acute attacks are extremely rare before puberty and most people affected will only have one or a few attacks in their entire life. However, people with VP 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 VP 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. Alternatively, 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 long periods with little food, can start acute attacks. It is important to have 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 VP should avoid oral contraceptives and HRTs that contain progesterone or progestogens. Injectable and implantable long-term