

Are there any particular places/times when I need to be careful with drugs?

Anaesthetics and surgical operations: You must tell your surgeon and anaesthetist in advance that you have porphyria. Safe local and general anaesthetics, sedatives, antibiotics and pain relief must be used.

Dentist: You must also tell your dentist that you have an acute porphyria. Dentists often use local anaesthetic agents and a safe one must be chosen.

Contraceptives and HRT: Women with acute porphyria should avoid oral contraceptives and HRT preparations containing progesterone or progestogens, if at all possible. Injectable and implantable long-term hormone preparations are very dangerous and must always be avoided.

In special circumstances, where risk is low and the benefits high, hormone-containing preparations may be considered (after discussion with your porphyria specialist or UKPMIS). Replacement doses, given through the skin from patches, are preferred as they can be rapidly removed.

What about immunisations/vaccines?

Childhood immunisations are safe, indeed it is vital to protect children from serious childhood diseases.

Vaccines for international travel are also generally considered safe. However, an acute attack has been reported following yellow fever vaccination so it is important to be cautious when using live vaccines in adults. A number of suitable antimalarial drugs are also available (see *UKPMIS SAFE list*).

The annual flu and pneumonia vaccines are also safe for those with an acute porphyria.

Topical preparations (e.g. gels, creams and lotions applied directly to the skin) including antifungals, lice treatments and eye drops are considered safe when applied to INTACT skin or mucosa.

Medic Alert/warning jewellery

Warning jewellery can be helpful in emergencies when you may not have an opportunity to explain that you have porphyria. Details on warning jewellery can be obtained from Medic Alert: www.medicalert.org.uk or 01908 951045.

Useful contact details

BPA telephone helpline: 0300 30 200 30

BPA email helpline: helpline@porphyria.org.uk

UK Porphyria Medicines Information Service (UKPMIS)

UKPMIS provides advice and a list of SAFE drugs for those with acute porphyrias: www.wmic.wales.nhs.uk/porphyria_info.php. UKPMIS can be contacted by telephone on 029 2074 3877 or 029 2074 2251.

European Porphyria Network: www.porphyria-europe.com

Rare Connect: www.rareconnect.org

Acute porphyria patient experience videos:
www.youtube.com/user/acuteporphyrias/videos

National Acute Porphyria Service (NAPS)

NAPS provides clinical advice and haem arginate where appropriate for patients having one-off acute attacks or recurrent attacks of porphyria. There are currently 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.

British Porphyria Association

Charity No: 1089609



Drugs and porphyria (acute porphyrias only) Information leaflet



Who does this leaflet apply to?

This leaflet **ONLY** applies to the acute porphyrias (*acute and mixed columns in the table below*) i.e. AIP, ADP, VP and HCP. **It does not apply to EPP, XLEPP, CEP or PCT.**

Why is it important to be careful with drugs in the acute porphyrias?

For those suffering from an acute porphyria (AIP, ADP, VP or HCP) attacks can be precipitated by drugs. Many medicines are capable of triggering acute attacks.

It is important to ALWAYS check the safety of any medicine or remedy with your doctor/pharmacist.

This includes prescription medicines as well as over-the-counter treatments, tonics and herbal remedies, some of which have been known to cause attacks. It is advised to avoid all herbal remedies unless you are sure of their safety.

While many medicines are considered suspect, there are many other drugs available and good alternatives can almost always be found.

- Even **people who have never had an acute attack** should be cautious when taking medicines. A combination of drugs with other factors such as stress, infection or poor nutrition may increase the risk of an acute attack.
- Though acute attacks are very rare before puberty, **it is also best if children take only safe drugs.**
- Also, it is very important for those who are **suspected of having an acute porphyria** (or are in the process of undergoing tests to eliminate the possibility) to be extremely cautious with drugs.

The response of people with porphyria to unsafe drugs is unpredictable. A reaction does not follow in every case. However, if there is a porphyria reaction, it will take the form of an acute attack, which normally develops within days of taking the unsafe drug.

Reactions such as dizziness, feeling faint, allergies or short-lived skin rashes (which may occur immediately or

very soon after taking the drug) are common after taking drugs and rarely have anything to do with porphyria.

Side effects of abdominal pain or sickness may occur with some medicines, but will not always indicate an acute attack. However, if this happens, it is important to contact your doctor/porphyria specialist.

How can I check the safety of a drug?

The UK Porphyria Medicines Information Service (UKPMIS, run by the Welsh Medicines Information Centre) has compiled a **list of SAFE drugs** for easy reference (*please see contact details overleaf*). **Acute patients should keep an up-to-date copy with them.** The safe list is updated annually using information collated internationally on drug safety over the previous year. This means that new drugs are sometimes added and at other times, drugs are removed following reports of adverse events.

Please contact UKPMIS to be added to their database to receive your new copy each year. There is also a link to the latest safe list on our BPA website. **Patients or their doctors can call UKPMIS for advice on drug options.** Drugs not on the safe list should only be taken after obtaining expert advice from UKPMIS or a porphyria specialist.

The European Porphyria Initiative (EPI) website www.porphyrria-europe.com contains further detailed information on prescribing in acute porphyria, including common prescribing problems (pain relief, anaesthesia, hormonal contraception).

Evidence about drug safety is drawn from three main sources: reports from doctors, medical professionals and patients; experimental systems (e.g. cell cultures); and drug metabolism data (how drugs are used in the body).

What if a safe drug cannot be found?

Some people with porphyria may occasionally need a drug, perhaps for a serious illness, that carries some risk of triggering an acute attack or which has been introduced so recently that there is little information about its effect on porphyria. In this situation, your doctor (after discussing the risks with a porphyria specialist) may decide to prescribe the drug for you with special follow-up.

When prescribing in acute porphyria, the benefit from using the ideal drug should always be assessed against the risk of provoking an acute attack, and the likely consequences of not using it. A drug should never be withheld if it is judged to be essential for optimum treatment (e.g. chemotherapy for cancer). Support in deciding the best alternative can be obtained by telephoning UKPMIS (*see overleaf*).

What if I have been taking an unsafe drug?

Some people may have used an unsafe drug without problems, even for many years, before being found – usually through family screening – to have latent porphyria. After discussion of the risk, these people may wish to continue on the drug and this is often allowed. However, if an additional drug needs to be prescribed, the possible risk of provoking an acute attack needs to be carefully considered and a change to a safer combination may be required. **Note:** even those who have taken an unsafe drug without problems in the past, could suffer an acute attack if they take it again.

All individuals starting therapy should be advised to report any adverse reaction (e.g. abdominal pain or dark urine), immediately.

Acute porphyrias	Mixed: acute porphyrias with skin symptoms	Cutaneous (skin) porphyrias - this information does not apply
Acute intermittent porphyria (AIP) ALA-dehydratase deficiency porphyria (ADP - also called plumboporphyria)	Variegate porphyria (VP) Hereditary coproporphyria (HCP)	Porphyria cutanea tarda (PCT) Erythropoietic protoporphyria (EPP) X-linked EPP (XLEPP) Congenital erythropoietic porphyria (CEP)