

What is porphyria?

Porphyria is a group of rare conditions that people are born with.

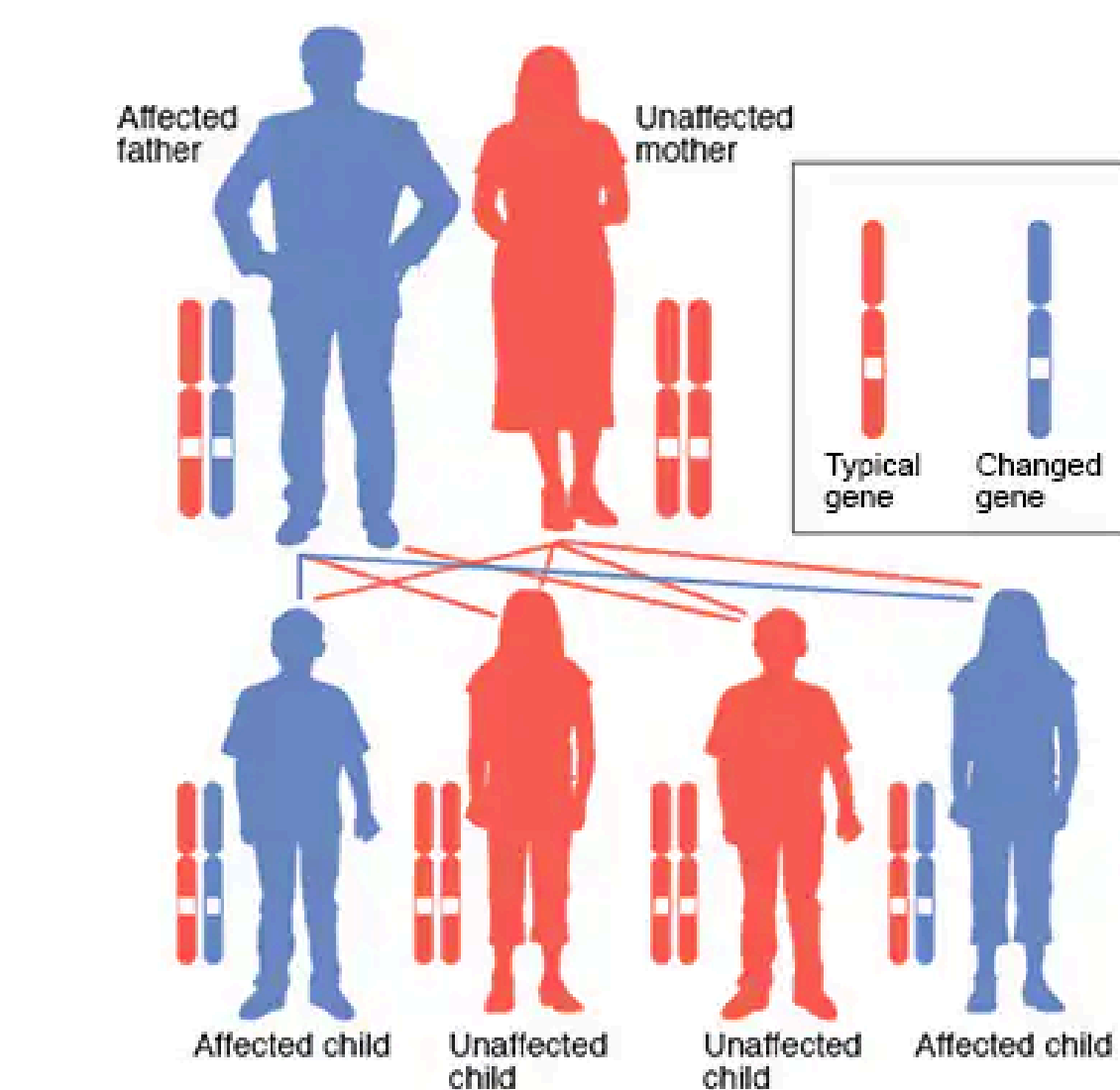
People with porphyria have trouble breaking down certain chemicals in their bodies. These chemicals can build up and cause illness.

There are two main kinds of porphyria:

- Acute (causes sudden attacks of sickness)
- Skin (causes skin problems)



What is acute intermittent porphyria (AIP)?



How do you get it?

Acute intermittent porphyria (AIP) is inherited, which means it can be passed from a parent to their child.

Each child has a 50% chance of getting the gene if one parent has it.

Boys and girls can get it equally.

Sometimes it looks like it skips a generation because some people never get symptoms.

Will it affect me?

Most people with AIP never have an attack.

Some people may have one or a few attacks in their life. A small number have more frequent attacks.

Attacks usually happen between the late teens and early forties. They almost never happen in children before puberty.

Even though attacks can be serious, most people get better with treatment.

What are AIP attacks like?

An AIP attack can make someone feel very unwell.

Common symptoms include:

- Severe pain, usually in the tummy, but sometimes in the back, arms or legs
- Feeling sick, being sick, or constipation (difficulty going to the toilet)
- Low salt levels in the blood
- A fast heartbeat or high blood pressure

Some people may also become confused during an attack.

Very rarely, someone might have seizures or their muscles may become weak, which can sometimes lead to paralysis (trouble moving parts of the body). These more serious problems can appear weeks after the attack starts.

Most AIP attacks last one to two weeks.

If someone gets muscle weakness or paralysis, they can recover, but it may take a long time.

What can cause (trigger) attacks?

Attacks often happen because of things that can be avoided, such as:

- Some medicines
- Alcohol
- Not eating enough or skipping meals
- Hormones (especially in women)
- Infections and stress



Medicines

People with AIP must be very careful with medicines, vitamins and herbal remedies. Some medicines are safe, and doctors have special lists to check. Medicines that are not on the safe list should only be taken after expert advice.

What else can cause attacks?

Alcohol

Alcohol is a very common trigger. The safest choice is not to drink alcohol at all. If someone does drink, it should be very small amounts.

Not eating enough or skipping meals

Going too long without food or eating very few calories can trigger an attack. It's important to have regular meals and a normal, balanced diet.

Hormones

Women are more likely than men to have attacks because of female hormones.

Some hormone medicines (like certain contraceptives which stop you getting pregnant) can trigger attacks and should usually be avoided.

Periods can sometimes trigger attacks, and doctors may help by giving treatments that stop periods for a while.

Pregnancy

Most pregnancies are safe, but there is a slightly higher chance of an attack. Doctors caring for a pregnant woman must know she has AIP.



What do I do if I get an attack?

Spotting an AIP attack early

It's important to notice an AIP attack as soon as possible so treatment can start quickly.

People who have had attacks before usually recognise the early signs.

If someone thinks an attack is starting, eating or drinking something sugary or starchy (like juice, sweets or bread) may help make the symptoms less serious.

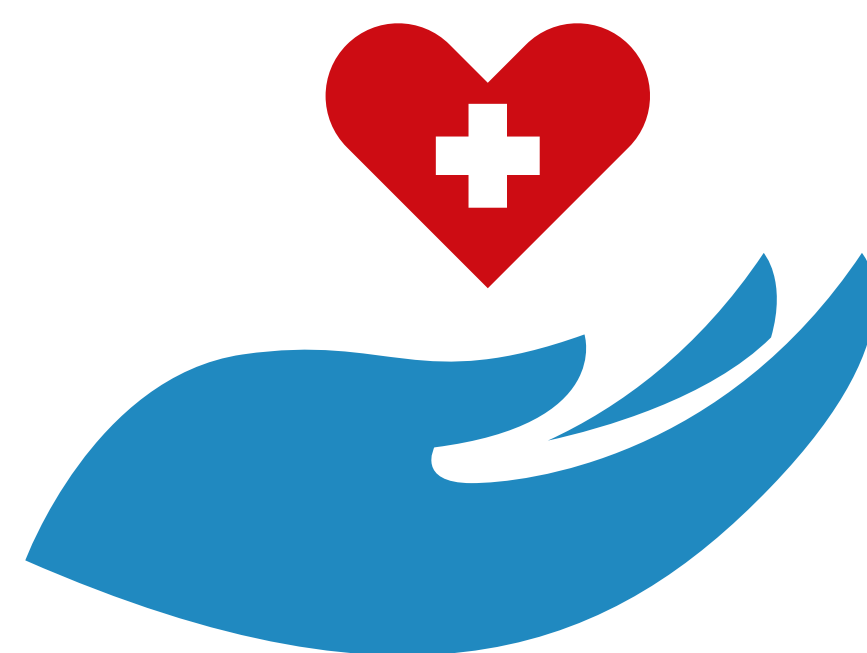
How do I know whether it is an attack?

It can be hard to tell what is or isn't an attack.

Many people with porphyria worry about tummy pain, but most of the time it is caused by normal problems, like:

- Stomach bugs
- Urine infections
- Bowel issues
- Appendicitis or other medical problems

Doctors can usually find out quickly by checking the urine for a chemical called PBG.



When to get help

If the symptoms are strong or do not get better within 24 hours, you should see a doctor. You might need to go to hospital, where doctors can:

- Test your urine to check if it's an AIP attack
- Start special treatment, such as haem arginate
- Help with symptoms like pain, sickness or not eating enough

Doctors can call the National Acute Porphyria Service (NAPS) for advice or treatment.

Hospital treatment

If someone needs to stay in hospital, they may be given haem arginate through a drip.

This helps the liver and reduces the body's need to make the chemicals that can trigger attacks.

Other treatments may include:

- Medicines for pain or sickness
- Medicines to help someone stay calm
- Extra calories through a drip or a feeding tube if they cannot eat normally

Living with AIP

Even though AIP can sound scary, most people with it live full, healthy lives.

Many never have an attack at all. And for those who do, attacks can usually be treated quickly, especially when they're spotted early.

By learning what triggers attacks and how to avoid them, people with AIP can stay well most of the time. Doctors and porphyria specialists are always there to help, and new treatments and research are making life even safer for people with acute porphyrias.

With good care, support, and understanding of the condition, most people with AIP can do the things they enjoy—go to work, have hobbies and live normal, happy lives.





Useful contact details

BPA telephone helpline: 0300 30 200 30

BPA email helpline: helpline@porphyria.org.uk

International Porphyria Network: porphyrianet.org

Medic Alert: medicalert.org.uk

YouTube video on AIP:

https://youtu.be/DpGg2seVQ_g



UK Porphyria Medicines Information Service (UKPMIS)

UKPMIS provides advice and a list of SAFE drugs which can be downloaded at wmic.wales.nhs.uk/specialist-services/drugs-in-porphyria. UKPMIS 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.

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: bipnet.org.uk.