

Using haem arginate (human hemin) in acute porphyria attacks

Supply

Established in 2012, the [National Acute Porphyria Service \(NAPS\) provides clinical advice, authorisation for use and supply of haem arginate treatment](#) 24 hours a day, seven days per week.

Authorised use of haem arginate within England, supplied via NAPS will be reimbursed through this service, which is funded by NHS England.

The service is provided in rotation from one of two [centres](#):

- King's College Hospital, London
- University Hospital of Wales, Cardiff

Indication

Haem arginate (Normosang®) is indicated to treat moderate to severe acute porphyria attacks and use should always be discussed with NAPS.

It is believed to be more efficacious if started early in an attack. Ideally within 12-24 hours after the diagnosis has been established, unless the attack is clinically mild and obviously resolving. It will not reverse an established neuropathy, although it may prevent its onset and may halt further progression of neuropathy if given sufficiently early.

Dose

The recommended dose is 3mg/kg (to a maximum of 250mg) once daily for 4 consecutive days.(5)

For convenience, 250mg (1 x 10ml ampoule) for adults may be given irrespective of their weight.(4,6) This would be suitable for patients weighing ≥ 70 kg. For patients weighing less consider rounding the dose to the nearest 25mg (1mL).

A course is not usually repeated except with advice from NAPS.(3) The effectiveness of long-term treatment has not been evaluated.(4,5)

Monitoring

Please refer to the [British and Irish Porphyria Network \(BIPNET\) guidelines](#).

Administration

Preparing a dose

Dilute the haem arginate immediately prior to administration, in sodium chloride 0.9% in a glass bottle. Use a plastic container (off-label use) if a glass bottle is not available to avoid delaying treatment in an emergency.

Giving a dose

Administer as an intravenous infusion in 100mL of sodium chloride 0.9% over 30 minutes into a large antecubital vein or preferably via a central line.

Use an in-line filter to filter out any unseen particles from the dark solution (most giving sets contain a 15 micron filter, but check packaging first).

Protect the product from light during the infusion.

After infusion, promptly flush the vein well with 100mL of sodium chloride 0.9%, initially as three or four 10mL boluses, and then the remaining volume may be infused over 10 to 15 minutes.

Symptoms generally start to improve within 24 hours and are usually significantly improved after 48 hours.

Cautions

The infusion is hypertonic and contains propylene glycol. Monitor the site during treatment as there is a risk of extravasation, which may cause skin discolouration.

Stop the infusion immediately if drug extravasation is suspected.

Haem arginate is irritant to the veins. Alternate arms each day to reduce the risk of inflammation and thrombophlebitis.

Problems with repeated use

Repetitive peripheral use may lead to loss of the superficial venous system and the consequent need for a central line. Central lines may also, in time, become obstructed with haem deposits.

Although unlicensed and lacking robust evidence, some anecdotal experience suggests administering haem arginate in 100mL of human albumin 20% to help reduce these problems and infusing over 60 minutes. Human albumin 20% is hyperosmotic, so if used, maintain adequate hydration and monitor electrolytes.

How it works

To produce haem, the body needs to convert porphyrin precursor chemicals ALA and PBG (5-aminolaevulinic acid and porphobilinogen) into more complicated substances called porphyrins. These are then converted from one type of porphyrin into another to form haem.

By replenishing hepatic haem stores the initial rate-limiting enzyme ALA synthase is inhibited by a process of negative feedback. The formation of porphyrins and their precursors, ALA and PBG, is reduced and symptoms improve.

After haem administration, urine ALA, PBG and porphyrins will fall within the first 24 hours.

Contact

University Hospital of Wales Switchboard 029 21847747

Ask for the National Acute Porphyria Service.

The switchboard can

- advise which centre is providing the emergency cover that week according to an agreed rota
- provide a telephone contact number or long-range pager number as arranged by each centre

During office hours, contact the patient's specific porphyria centre (Cardiff/King's) for advice.

Useful Links

[Normosang® Summary of Product Characteristics](#)

[Injectable Medicines Guide](#)

[British and Irish Porphyria Network \(BIPNET\)](#)

[British Porphyria Association \(BPA\)](#)

[A list of medicines that are safe in the acute porphyrias](#) is produced annually by the UK Porphyria Medicines Information Service (UKPMIS). They can also be contacted for specialist porphyria advice on choosing medicines, on telephone 029 21842251. This service is available to healthcare professionals and people with porphyria in the UK.

Mae'r daflen hon hefyd ar gael yn Gymraeg. This leaflet is also available in Welsh.