Porphyria - Summary Clinical Guideline

Reference no.: CG-GASTRO/2018/018

Acute Porphyria Emergency Management Guidelines

Acute intermittent porphyria (AIP), Variegate porphyria (VP) and Hereditary Coproporphyria (HCP) are autosomal dominant conditions, which may lead to potentially life threatening acute neurovisceral attacks.

Attacks are uncommon but may be precipitated by

- UNSAFE prescribed or illicit drugs
- Infection
- Stress
- Excess alcohol
- Reduction in calorie intake
- Sex hormone fluctuation (especially raised progesterone in luteal phase of menstrual cycle)

Clinical features during an attack

- Abdominal pain - Severe, poorly localised No evidence of acute abdomen on examinations. Level of pain appears out of keeping with physical signs and requires large doses of opiate administration.

- Nausea/vomiting/constipation
- Hypertension and tachycardia and (rarely) arrhythmias
- Convulsions - Frequently associated with hyponatraemia
- Peripheral motor neuropathy - May progress to flaccid paralysis, respiratory insufficiency, difficulty swallowing, urinary retention or incontinence
- Psychiatric symptoms such as agitation, insomnia, confusion, psychosis

MANAGEMENT
Clinical Assessment

- Ensure adequate analgesia – usually opiates
- Discontinue any un-safe drugs
- Monitor pulse and blood pressure, at least 4 hourly
- Check motor power and ventilator function. Evidence of respiratory insufficiency
- Requires immediate transfer to ITU
- **Consider and exclude other causes of abdominal pain**

Biochemical assessment

- U+E (daily or more frequently in hyponataemia)
- FBC, Ca, Mg, LFTs and CRP on presentation then twice weekly
- Urine porphobilogen (PBG) testing may MAY be helpful. Collect 10 mL random urine in a plain tube **(protect from light)** and send to laboratory for porphyria screen.
- This sample should be collected PRIOR to starting any Haem arginate.

Supportive Treatment

Indication	Suggested safe drugs
Maintain fluid/calorie intake a) Tolerating oral intake	25% Oral glucose solution
Maintain fluid/calorie intake b) Not tolerating oral intake	IV 0.9% sodium chloride containing 5% glucose AVOID IV glucose in water solution including glucose 5% and 10% as may aggravate hyponatraemia.
Pain	Paracetamol, Morphine, Diamorphine and Fentanyl. AVOID PETHEDINE
Vomiting	Promazine, Prochloperazine or Ondansetraon
Agitation/Anxiety	Chlorpromazine
Hypertension/tachycardia	Atenolol, Propranolol or Labetalol
Convulsions	Diazepam, Clonazepam or Magnesium Sulphate

Specific treatment:

Haematin (Haem-arginate; Norosang)

SUPPLIED BY THE NATIONAL ACUTE PORPHYRIA SERVICE (25 mg/mL stock solution**)**

- This should she administered as soon as possible on diagnosis of an acute attack of porphyria.
- Dose: 3 mg/kg once daily (maximum 250 mg daily) for 4 days
- If response inadequate, consider further 4 day course
- Drug should be diluted immediately prior to use in 100 mls of 0.9% Sodium Chloride or 20% human serum albumin and infused with a one hour (maximum rate 2 mLs per minute)
- After infusion: Flush vein with 250ml Sodium Chloride (0.9%) initially 3 – 4 boluses of 10ml, the remainder under gravity.
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Further information

CLINICAL ADVICE and Haem Arginate supply: British and Irish Porphyria Network: T: 02920 747747 (24/7 service) www.bipnet.org.uk