BIMDG
British Inherited Metabolic Disease Group

ADULT EMERGENCY MANAGEMENT
ACUTE PORPHYRIA

Patient name:	**Date of birth:**
Diagnosis:	
Hospital	
Telephone:	

IMMEDIATE ACTIONS

For patients in England, Scotland and Wales, please contact the National Acute Porphyria Service (NAPS) for advice:

Within working hours: phone the NAPS team responsible for treating the patient if known. This will be either: King's College Hospital London (020 3299 5776) or University Hospital of Wales Cardiff (029 20746588)

Out of working hours (or new patients at any time): An emergency service is provided 24 hours a day and 7 days a week. Phone the switchboard of the University Hospital of Wales on 02920 747747 who will provide contact details for the on-call porphyria team.

BACKGROUND:

The acute porphyrias (acute intermittent porphyria, variegate porphyria, hereditary coproporphyria) are autosomal dominant conditions. Patients are at risk of acute neurovisceral attacks, which may be life threatening. Acute attacks are uncommon and do not affect all those who inherit the condition. It may be difficult to distinguish between an acute attack and other causes of acute abdominal pain. Acute attacks may be precipitated by infection, stress, unsafe prescribed or illicit drugs, dieting or excess alcohol consumption. Multiple factors may be involved or there may be no obvious precipitating event. The symptoms and signs are due to neuronal dysfunction affecting the autonomic, and/or motor and/or central nervous system.

CLINICAL FEATURES:

An acute attack of porphyria almost invariably starts with constant abdominal pain, which becomes progressively more severe and is associated with nausea, vomiting and constipation. The patient may also complain of pain in the lower back, buttocks and thighs. The abdominal pain is diffuse with no localising signs or evidence of an acute abdomen on examination. The level of pain appears out of keeping with the physical signs and requires large doses of parenteral opiate administration for effective relief. This may require support from the pain team.

Other symptoms and signs include:

Hypertension and tachycardia.
Seizures, may be related to hyponatraemia
Muscle weakness, may progress to flaccid paralysis and respiratory insufficiency
Psychiatric symptoms, such as agitation, insomnia, confusion, psychosis

The current list of drugs considered safe to use in acute porphyria can be found at:
https://www.wmic.wales.nhs.uk/specialist-services/drugs-in-porphyria/

Detailed information on diagnosis and treatment of acute porphyria is available at:
www.porphyria-europe.org

RECOMMENDED MANAGEMENT

CLINICAL ASSESSMENT

Assess general condition and analgesic requirement
Monitor pulse and blood pressure
Check motor power and ventilatory function
Review all prescribed medication
Consider and remove possible precipitating factors (unsafe drugs, alcohol fasting, infection)
Consider and exclude other causes of abdominal pain

INITIAL INVESTIGATIONS

1. Urea & electrolytes - hyponatraemia is common
2. Full blood count
3. Urine porphobilinogen - collect a 10ml random urine sample in a plain labelled tube and protect from light prior to analysis. Sample should be collected before starting haem arginate. In known patients treatment should not be delayed while waiting for the result.
4. Other tests as indicated by clinical findings eg CRP, infection screen

TREATMENT:

1. Analgesia – Opiates are almost always indicated, and should be administered regularly. Consider a morphine PCA if pain is severe. Dihydrocodeine and morphine are safe. Pethidine should be avoided.
2. Maintain fluid and calorie intake
    a. If tolerating oral intake – encourage carbohydrate
    b. If not tolerating oral intake – start intravenous 0.9% sodium chloride containing 5% glucose (maintenance 2 litres per day). Avoid intravenous glucose in water solution, including dextrose 5% and 10%.
3. Symptomatic treatment as indicated using medication that is safe in porphyria such as:
    a. Vomiting - ondansetron, cyclizine, prochloperazine
    b. Agitation - chlorpromazine
    c. Seizures - diazepam, clonazepam, magnesium sulphate
    d. Hypertension/tachycardia - propranolol, labetalol
    e. Constipation - bulk laxatives, senna
4. Haematin (haem arginate; Normosang®) is indicated if any of the following present: severe persistent pain, persistent vomiting, neuropathy, seizures, hyponatraemia, psychosis, severe hypertension and tachycardia

- Intravenous haem arginate (Normosang, Orphan Europe, UK) is available as stock solution at 25 mg/ml. This is supplied free of charge through the National Acute Porphyria Service for all patients living in England and Scotland. For Welsh patients contact the Cardiff NAPS service.
- Dose: 3 mg/kg daily (up to maximum of 1 vial/250 mg daily) on 4 consecutive days
- Dilute immediately before use in 100 ml 0.9% sodium chloride solution and protect from light
- Infuse intravenously over 30-60 min via large antecubital vein (or central line) using a giving set with a 15-20 micron inline filter
- Flush vein with 250 ml 0.9% sodium chloride solution (initially 3-4 boluses of 10ml, then remainder under gravity).
- Vascular complications may be limited by diluting haem arginate in 20% human serum albumin and/or alternating arms for consecutive infusions
- Side effects are minimal apart from thrombophlebitis at site of administration

Last reviewed December 2017