St Helens and Knowsley Teaching Hospitals NHS
NHS Trust

Emergency Management of Patients with Acute Porphyria v1.1

Step 1: Assess

Patients with known Acute Intermittent Porphyria (AIP) require a comprehensive history and examination; symptoms can become life threatening if not managed properly. Patients will often be familiar with the type of abdominal pain/other symptoms that usually herald an acute attack. Ask about precipitants: drugs, periods, alcohol, fasting, stress, infection. Consideration alternative diagnoses such as urinary tract/chest infection and appendicitis should always be considered by appropriate testing. Look for neuromuscular weakness causing respiratory failure or hyponatraemia (consider ICU) vomiting and depression.

Step 2: Ring National Acute Porphyria Service (NAPS) on 029 20747747

Following initial assessment, ring NAPS (24 hour service), once you have relevant clinical and lab results. They will guide further management including haemarginate.

Step 3: Get an urgent urine sample

Even in patients with known AIP, you must send off an immediate patient urine sample for Porphobilinogen (PBG) levels *before* starting haemarginate (OCS>Request entry>Biochemistry>Urine>Random Urine>Porphyria) in a plain universal container shielded from light, clearly labelled with the patient name, date of birth and date. Alert biochemistry (1832)-they the sample off to NAPS for testing.

Step 4: Prescribing

Before prescribing and giving ANY medication, check that the drugs are safe using the Cardiff safe drug list or www.wmic.wales.nhs.uk if the direct link is broken.

Step 4: Specific drug management

NAPS will guide as to whether haemarginate should be given. If directed, use a large vein cannula. The dose is 3 mg/kg once daily (max. 250 mg daily) for 4 days initially, -NAPS will advise. Stock held in box in fridge in the emergency drug room on the 5th floor or pharmacy. Ask the MET team to access if the patient has a portacath. If vomiting give 0.9% normal saline (not 10% dextrose-hyponatraemia risk). If severe hyponatraemia (<120mmol/l), click here for GAIN guideline

The pain from acute intermittent porphyria can be extremely distressing. Do not accuse these patients of malingering -the pain is real and patients may need much higher than usual doses of intravenous morphine (avoid oral opiates or tramadol) to control acute abdominal pain. Consider expert advice from ICU if severe hyponatraema or respiratory failure, the acute/chronic pain team and a dietician for improving calorie intake. Click here for further detailed best practice guidance.

Step 5: Check EDMS for an inpatient care plan-some patients may have this in place. NAPS should advise re duration of haemarginate dosing

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