Worcestershire Acute Hospitals NHS Trust

# Haemolytic Uraemic Syndrome (PIP)

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## The following guidance is taken from the Partners In Paediatrics (PIP)

# HAEMOLYTIC URAEMIC SYNDROME

## RECOGNITION AND ASSESSMENT

**Definition**

- Triad of features
- microangiopathic haemolytic anaemia
- thrombocytopenia
- acute kidney injury (AKI)

**Symptoms and signs**

- Diarrhoea with blood and mucus (rarely haemolytic uraemic syndrome can occur in absence of diarrhoea), rectal prolapse
- dehydration if diarrhoea has been severe (see **Diarrhoea and vomiting** guideline)
- check BP: hypotension
- Vomiting
- Abdominal pain
- Pallor, lethargy
- Reduced urine output/facial puffiness
- Tachycardia
- Reduced consciousness: consider cerebral oedema, intracranial thrombosis/haemorrhage
- Seizures: consider hyponatraemia, cerebral oedema, intracranial thrombosis/haemorrhage
- Paralysis: consider intracranial thrombosis/haemorrhage
- Over-hydration
- oedema (periorbital/pedal) variable
- weight gain, observe trend
- raised jugular venous pressure (JVP) indicates volume overload
- oliguria (urine output <1 mL/kg/hr)
- tachypnoea
- liver enlargement
- Non renal complications:
- toxic megacolon
- perforation
- intussusception
- rectal prolapse
- cardiomyopathy
- diabetes mellitus
- intracranial thrombosis, haemorrhage, oedema

**Investigations**

- FBC and blood film (look for fragmented red cells)
- low Hb and platelets
- Clotting studies (normally activated – should not be DIC picture)
- U&E, creatinine, LDH (to confirm haemolysis)
- Bicarbonate
- Calcium, phosphate, uric acid
- Glucose, amylase
- Liver function tests
- Serum *E. coli* O157 lipopolysaccharides (LPS) antibodies
- Urine stick test for significant blood and protein (indicating glomerular damage) and leucocytes
- Stool culture for *E. coli* (and typing for O157 strain)

# IMMEDIATE TREATMENT

- Admit, discuss with regional paediatric nephrology team in all cases
- Strict fluid balance, electrolyte monitoring and management, see Acute kidney injury guideline
- Dehydration
  - if signs of hypovolaemic shock give circulatory support (sodium chloride 0.9% 20 mL/kg IV immediately)
  - correct dehydration (see Diarrhoea and vomiting guideline)
- Over-hydration
  - if signs of overload/cardiac failure, furosemide 2–4 mg/kg (commence at 2 mg/kg and adjust to response) IV over 1 hr (maximum rate 4 mg/min), repeated 6-hrly if response obtained
  - if furosemide ineffective, discuss dialysis with regional paediatric renal centre
- Hypertension (see **Hypertension** guideline)
- Anaemia
  - daily FBC: only transfuse after discussion with regional paediatric nephrology team as may require dialysis. If asymptomatic, Hb can drop as low as 60 g/L
- Thrombocytopenia
  - do not transfuse platelets unless there are life-threatening bleeds/instrumentation required
- AVOID antibiotics, anti-diarrhoeal treatment, NSAIDs, and other nephrotoxic medication
- Observe for non-renal complications e.g. encephalopathy and seizures, cardiomyopathy, diabetes mellitus (twice daily BM sticks for the first 48 hr)
- Protein and sodium restriction

**Tertiary referral**

- If significant renal impairment (oligo/anuria, rising creatinine, severe acidosis, hyperkalaemia or complications) dialysis required (see **Acute kidney injury** guideline), refer to regional paediatric nephrology team
- Refer urgently if non-diarrhoeal haemolytic uraemic syndrome

# DISCHARGE FROM HOSPITAL

- Patient may be discharged when all following criteria met:
  - diarrhoea/abdominal pain resolved
  - Hb stable (haemolysis ceased)
  - drinking fluids freely and passing normal amounts of urine
  - urea and electrolytes improving with normal serum potassium
- Prescribe folic acid 2.5 or 5 mg daily until Hb normal

**Follow-up**

- Weekly until renal function normal
  - if impaired renal function or proteinuria persists, arrange paediatric renal follow-up
- Once renal function normal, arrange GP or general paediatric follow-up every year to check BP and early morning urine (protein:creatinine ratio) with a detailed renal specialist review every 5 yr for formal GFR
- Advise that women with history of haemolytic uraemic syndrome require close monitoring during pregnancy
- Advise about avoiding smoking and obesity

# DISCHARGE FROM FOLLOW-UP

- Renal function normal
- No proteinuria
- Renal growth and function satisfactory at 5-yrly review for 15 yr