Princess Margaret Hospital for Children Emergency Department Guideline

PAEDIATRIC ACUTE CARE GUIDELINE			
Haemolytic Uraemic Syndrome			
Scope (Staff):	All Emergency Department Clinicians		
Scope (Area):	Emergency Department		

This document should be read in conjunction with this DISCLAIMER http://kidshealthwa.com/about/disclaimer/

Haemolytic Uraemic Syndrome

Haemolytic Uraemic Syndrome (HUS) is a disease characterised by:

- Microangiopathic haemolytic anaemia (destruction of red blood cells)
- · Acute renal failure
- Thrombocytopaenia

General

Typical Heamolytic Uraemic Syndrome

- Is among the commonest causes of acute renal failure in children
- Mortality of 5-10%
- In the majority of cases it follows gastrointestinal tract infection with verotoxin producing *E. coli*, but it also can be induced by *Shigella*, *Campylobacter* and different viruses. In most of these there is preceding diarrhoea, which is frequently bloody (D+HUS)

Atypical Heamolytic Uraemic Syndrome

- Represents in 5-10% of cases
- May develop without a diarrhoea prodrome
- Causes:
 - Complement defects
 - Familial cases
 - Drug toxicity (e.g. chemotherapy, Tacrolimus, Cyclosporin, oral contraceptives, Valacyclovir)
 - Immune mediated (e.g. Quinine, Clopidogrel)
 - Malignancy
 - Hereditary (e.g. inborn error of cobalamin deficiency)
 - Connective tissue disease (e.g. SLE, scleroderma, antiphospholipid antibody syndrome)
 - Other glomerunephritides (e.g. APIGN, memobranoproliferative GN)

History

- Most children with HUS present 5-10 days after the onset of a bloody diarrhoea with:
 - Oliguria
 - Haematuria
 - Anaemia
 - o Oedema
 - Renal failure
 - Hypertension
- Myocardial infarction, stroke, pancreatitis, liver necrosis, encephalopathy and seizure have also been described

Examination

Full systems examination

Assessment of cardiovascular and intravascular volume status is very important

Consider:

- Thirst, restlessness and confusion
- Capillary refill
- Skin turgor
- Olyguria
- Fontanelle tension
- Blood pressure
- Heart rate
- Evidence of oedema

Investigations

• FBC, film and differential

- U&E
- · Liver function tests
- CRP
- BGL
- Coagulation profile
- Group and hold
- E-coli serology (clotted sample red top tube)
- Blood cultures if Pneumococcal cause suspected
- Stool M, C & S
- Urinalysis and urine M, C & S
- Other investigations as clinically indicated (e.g. CXR, renal US, ECG, head CT or MRI, EEG)

Differential diagnoses

- Sepsis
- Acute post-Streptococcal glomerulonephritis
- Disseminated intravascular coagulation
- Immune thrombocytopaenic purpura (ITP)
- Thrombotic thrombocytopaenia purpura
- Systemic lupus erythematosis (SLE)
- Vasculitis

Initial management

Fluid Management:

- Establish IV access
- Hypovolaemia should be treated with 0.9% saline boluses, or packed red blood cells, depending on the clinical status (usually 0.9% saline in the ED setting)
- In the presence of oligo-anuria and fluid overload, fluid should be administered cautiously and should not exceed insensible fluid losses plus urine output (or less)
 - Insensible fluids:
 - 0-10kg weight give 25mL/kg/day
 - 10-20kg weight give 12.5mL/kg/day
 - Then 5mL/kg/day for each additional kg over 20kg weight
 - Hyponatremia is treated with fluid restriction
 - Electrolyte abnormalities: please discuss with Paediatric Renal Team

Altered consciousness / focal neurological signs:

• If these develop, immediate discussion should be undertaken with a senior colleague (e.g. ED Consultant, Renal Consultant or PICU Consultant)

Hypertension:

- Usually secondary to volume overload
- If unresponsive to diuretics, try vasodilator treatment (e.g. Nifedipine)

Abdominal pain and vomiting:

- Due to colitis in post-diarrhoeal HUS
 - Treat initially with paracetamol
- May require opiate analgesia (but this will desrease bowel motility and should be avoided if possible)
 - Do not prescribe NSAIDs such as ibuprofen

Antibiotics

• Indicated in pneumonia related HUS (not in typical D+HUS)

Further management

Indications for Dialysis:

- Fluid overload resistant to diuretic therapy
- Hyperkalaemia
- Intractable acidosis
- Symptoms of uraemia
- Likely progression of one of the above

Patients needing dialysis (most), will usually be placed on peritoneal dialysis (PD). Exceptions are those with severe colitis, cerebral HUS or profound metabolic abnormalities (where haemodialysis or haemofiltration techniques may be considered)

All patients diagnosed with HUS need to be admitted

The Paediatric Renal Team (and General Surgical Team for PD catheter / line placement) should be consulted early

Tags

abdominal, acute renal failure, bloody stool, diarrhoea, e coli, haemolytic uraemic syndrome, hus, oedema, oliguria, renal, stool, thirst

This document can be made available in alternative formats on request for a person with a disability.

File Path:					
Document Owner:	Dr Meredith Borland HoD, PMH Emergency Department				
Reviewer / Team:	Kids Health WA Guidelines Team				
Date First Issued:	6 June, 2014	Version:			

Last Reviewed:	9 October, 2017	Review Date:	9 October, 2020	
Approved by:	Dr Meredith Borland	Date:	9 October, 2017	
Endorsed by:	Medical Advisory Committee	Date:	9 October, 2017	
Standards Applicable:	NSQHS Standards:			

Printed or personally saved electronic copies of this document are considered uncontrolled