

Australian Paediatric Surveillance Unit

PROTOCOL - HAEMOLYTIC URAEMIC SYNDROME

Objectives: To study the epidemiology of Haemolytic Uraemic Syndrome in Australia; to relate its incidence to geographical location, age, sex and race; to elicit clinical features associated with HUS, in particular its association with or without a diarrhoea prodrome; to identify whether relapsing HUS occurs in Australia and; to determine the role of Shigatoxin-producing enterohaemorrhagic *E.coli* in HUS associated with diarrhoea.

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Summary protocol:

The Haemolytic Uraemic Syndromes are a heterogeneous group of disorders characterised by microangiopathic haemolytic anaemia (anaemia with microscopic evidence of fragmented red blood cells), thrombocytopenia and acute renal impairment (oliguria or anuria) with elevated serum urea and creatinine).

Two distinct clinical sub-groups have been identified. In the first there is a prodrome of diarrhoea (usually bloody), disease occurs in the summer and the majority of patients make a good renal recovery. In the second there is no diarrhoeal prodrome or seasonal variation. However HUS may be family history of Haemolytic Uraemic Syndrome, may be precipitated by infection eg *Streptococcus pneumoniae* disease or disease may be chronic relapsing. Outcome is thought to be worse in this group.

Shigatoxin producing Enterohaemorrhagic *E.coli* (STEC) have been implicated in the pathogenesis of HUS associated with diarrhoeal. STEC of the serotype O157:H7 are most frequently implicated in North America, Europe and Japan. APSU study to date suggests that O157:H7 is rare in Australia, where O111:H- recently caused a large "outbreak" and is the most common stool isolate in sporadic cases. No National figures have been previously available in Australia regarding the incidence of HUS. APSU figures estimate a minimum incidence of 0.58/100,000 for children <15 years (1.27 <5years). New information on virulence characteristics of STEC isolates in Australia has also been obtained.

CASE DEFINITION:

Any child less than 16 years of age with Haemolytic Uraemic Syndrome, defined as:

1. **Microangiopathic haemolytic anaemia Hb <10g/dl with microscopic evidence of fragmented red blood cells)**
2. **Thrombocytopenia (Platelets < 100,000 x 10⁹) and**
3. **Acute renal impairment (oliguria or anuria with elevated serum urea a creatinine)**

N.B. There may or may not be a history of preceding diarrhoeal illness. The degree of anaemia, thrombocytopenia renal impairment may vary considerably (some children having minimal haematological disturbance in the presence of significant renal impairment) and they may not all be present simultaneously. Septicaemia, chronic renal failure, collagen or vascular disorders and malignant hypertension may have a similar presentation and should be excluded.

Reporting instructions:

Please report any new patient seen in the last month who fulfils the case definition for HUS and remember to

1. Notify on monthly card
2. Send stool and /or serum specimens to MDU (see below)
3. Notify the relevant public health authority

Additional instructions regarding collection of biological specimens:

1. Phone **03 8344 5713** or Fax **03 8344 7833** (Microbiological Diagnostics Unit) to request a pre-paid transport container. (This will include the container shippers declaration, instructions **Prepaid** consignment note.) Page Dr Geoff Hogg **96252689** if you wish to discuss

2. Collect specimens:

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| | i) | Stool sample at 4°C |
| and/or | ii) | Rectal swab in Stuart's transport medium |
| and | iii) | Paired serum samples |
| and | iv) | <i>E.coli</i> isolates from faeces or implicated food (including <i>E.coli</i> 0111, 0157, 0113, 026) if available |

Note: Faecal samples should be obtained as soon as possible after diagnosis of HUS.
Sera should be acute and convalescent (approximately 2 weeks apart)
Faeces will be examined for verotoxin and VTEC
VTEC isolates will be characterised and stored.
Sera will aid assessment of antibody response.

3. Label specimens as follows:

- i) Attention Dr Hogg: HUS Study
- ii) Name and address of reporting clinicians
- iii) The first two letters only of the patients first name and surname
- iv) Patient's date of birth and postcode.

4. When the specimen is ready to send and air transport is required **ring the Microbiological Diagnostics Unit for instructions on air freight provision**

Samples should be addressed to:
Dr Geoff Hogg
Microbiological Diagnostic Unit
Department of Microbiology
University of Melbourne
Parkville VIC 3052

NB This laboratory (MDU) administers the National Enteric Pathogens Surveillance Scheme (which includes EHEC isolates). If EHEC is detected in a stool specimen MDU will notify the reporting clinicians and the appropriate public health authority.