

CQ_IBK_aHUS_01 / version 25/11/09

European Paediatric Research Group for HUS and related disorders

Case questionnaire for diarrhoea negative/VTEC (STEC) negative cases – acute phase

1. INSTRUCTIONS

Please type within the spaces indicated. Missing data should be left blank. To create a pedigree, symbols can be copied from the worked example in footnote 4. The declaration of consent must be affirmed in every case. The questionnaire can be returned electronically.

2. DEFINITION OF HUS

- microangiopathic hemolytic anemia: hemoglobin < 10g/dl with fragmented erythrocytes
- thrombocytopenia: platelet count < 130 000/mm³
- renal impairment: serum creatinine > age related range or GFR < 80 ml/min/1.73m² by Schwartz Formula. (Note renal impairment may not be evident at onset, investigator to use discretion in this circumstance)

3. EXCLUSION

- single episode of HUS preceded by diarrhoea
- single episode of HUS without diarrhoea but with evidence of VTEC infection
- disseminated consumptive intravascular coagulation, eg secondary to sepsis

4. DECLARATION

The local investigator confirms that the patient and/or the patient's parents/guardians have given permission for this information to be used by the European Paediatric Research Group for HUS, and a record of such agreement is retained in the patients medical record.

Confirmed:

5. IDENTIFICATION OF PHYSICIANS: LOCAL INVESTIGATOR

Name: _____ Surname: _____
Unit/Department: _____
Hospital: _____
Address: _____
Tel: _____ Fax: _____
E-mail: _____

Date of filling out: _____

6. IDENTIFICATION OF PATIENT

Date of birth: ___/___/___

Hospital: _____

Sex: Female Male

first letter of name:

first letter of surname:

7. CLINICAL EVENTS IN MONTH BEFORE DIAGNOSIS:

no yes If yes, date of onset (DD/MM/YY) ___/___/___

Diarrhoea	no	yes
Bloody diarrhoea	no	yes
Abdominal pain	no	yes
Vomiting	no	yes
Fever (T>38°C)	no	yes
Respiratory infection	no	yes
Others	no	yes

If yes, specify _____

7.1.DRUGS:

Given in month before onset: no yes

If yes,
specify _____

8.DESCRPTION OF ACUTE PHASE

Date of diagnosis of first episode of HUS (DD/MM/YY) ___/___/___

8.1. RENAL IMPAIRMENT

Oligo-anuria (<10ml/kg/24h)	no	yes	if yes, duration (days)
Dialysis necessary	no	yes	if yes, duration (days)
Maximum serum creatinine (before dialysis)			mg/dl
		µmol/l	
Creatinine clearance (Schwartz formula) ³			ml/min/1.73 m ²

8.2. HEMATOLOGICAL DATA

Minimum hemoglobin level g/dl

Schistocytes seen	no		yes	if yes	%
Minimum thrombocyte count		x10 ³ /mm ³			
Neutrophil count at diagnosis		x10 ³ /mm ³			
Minimum haptoglobin level		units	normal value		
Maximum LDH level		units	normal value		

8.3. HYPERTENSION Score (see footnote 1):

8.4. CNS INVOLVEMENT no yes

If yes, give details and include any imaging or relevant investigations:

8.5. GASTROINTESTINAL INVOLVEMENT no yes

If yes, give details:

8.6. PANCREATIC INVOLVEMENT INCLUDING DIABETES no yes

If yes, give details:

8.7. CARDIAC INVOLVEMENT

Cardiomyopathy: no yes

If yes, give details: e.g. Echocardiogram findings

8.8. OTHER SYSTEM INVOLVEMENT no yes

If yes, give details:

9. RELAPSING HUS

Relapse is defined as a return to microangiopathic hemolytic anemia, and/or thrombocytopenia and/or renal impairment at least 2 weeks after remission of HUS. Remission of HUS is defined as regaining a hemoglobin >10g/dl without fragmented red cells, and normal platelet count > 130 000/mm³.

yes unknown or 1st episode

If yes, please fill in:

(Outcome see footnotes 1,2 and 3)

Relapse No	Date	BP score	Proteinuria score	Creatinine clearance (Schwartz formula)
1				
2				
3				
4				
5				
6				
7				

10. FAMILIAL HUS no yes

If yes, give details:

Member: Description: Outcome:

DNA obtained from immediate family members no yes

11. TREATMENT

Please complete this page for the 1st episode, and duplicate it for each relapse. To make it clear which episode is being described please indicate:

First episode or relapse number: _____

Packed RBC	no	yes	If yes, number of units
Platelets infusion	no	yes	If yes, number of units
Plasma infusions	no	yes	
If yes, volume per infusions		ml/kg	number of infusions
Plasma exchanges	no	yes	
If yes, volume exchanged per session		ml/kg	number of sessions
Replacement by albumin	no	yes	
Replacement by plasma	no	yes	
Plasma cryosupernatant	no	yes	
IV immunoglobulins	no	yes	
If yes, dose per infusion		ml/kg	number of infusions
Antiplatelets agents	no	yes	
Heparin	no	yes	
Others	no	yes	

Therapy after remission? Yes No

If yes, please specify:

12. INVESTIGATION TOWARDS AETIOLOGY

12.1. Stools:

Positive for O157:H7	no	yes	unknown, not done
Positive for another VTEC	no	yes	unknown, not done
VT gene positive in stools	no	yes	unknown, not done
Positive for another bacteria	no	yes	unknown, not done

If yes, please specify _____

12.2. Serum:

Anti VTEC LPS IgM positive	No	yes	unknown, not done
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If yes, please specify serotype

12.3. Evidence of other infective causes no yes_

If yes, please specify _____

12.4. Complement

	Value/units	Normal range	Date of sample (DD/MM/YY)
CH 50			
APH50			
C3			
C3d			
C4			
Factor H protein			
Factor B			
Factor I			
MCP			
Faktor H Antikörper			

Factor H function and gene analysis samples sent to:

Results:

Other complement abnormality	no	yes
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If yes, please specify

12.5. Von Willebrand factor protease

vWF protease activity, samples sent to: _____

Result: Activity % Anti vWF protease antibodies no yes

12.6. further auto antibodies detected/suspected?

Endomysiale Antibodies (EMA) : _____

Transglutaminase-Antibodies (TGA): _____

Antigliadin-Antibodies (IgG-AGA, IgA-AGA): _____

Thyreoidea-Receptor-Antibodies (TRAK): _____

Anti-Neutrophile cytoplasmatic Antibodiesr (ANCA): _____

Other: _____

13. RENAL HISTOLOGY Documented no yes

If yes,

Date of renal specimen collection (DD/MM/YY) ___/___/___

Number of glomeruli per specimen

Results:

Predominant glomerular thrombotic micro-angiopathy (TMA)

Predominant arterial/arteriolar TMA

Cortical necrosis:

minimal

moderate/patchy

diffuse/extensive

FOOTNOTES

Footnote 1:

Blood pressure/ Hypertension:

Hypertension: Systolic and/or diastolic BP>97th percentile

According to local reference values while not on antihypertensive therapy

Score:

0=	Normal BP \leq 95 percentile for sex and height	
1=	Mild hypertension:	< 10mmHg over 95 th percentile
2=	Moderate hypertension:	10-30mmHg over 95 th percentile
3=	Severe hypertension:	>30mmHg over 95 th percentile

Footnote 2:

Proteinuria:

Score:

0=	Albustix (or equivalent) trace or negative on early morning urine sample or protein/creatinine <20mg/mmol or <0.2g/g
1=	Mild to moderate proteinuria, Albustix 1+ to 2+ (=up to 1g/l), or protein/creatinine ratio 20-200 mg/mmol or 0.2-2.0g/g
2=	Heavy proteinuria, Albustix 3+ or 4+, or protein/creatinine > 200mg/mmol or >2.0g/g

Footnote 3:

Creatinine Clearance according to Schwartz formula:

Clearance (ml/min/1.73m²) = $\frac{Ht (cms) \times k}{Creatinine (micromol/l)}$

Creatinine (micromol/l)

to convert creatinine in mg/dl to micromol/l multiply by 0.885

k values

Premature infant	29
0-2 years	40
2-12 years	48
13-21 years (girls)	48
13-21 years (boys)	62