

CQ_IBK_aHUS_02 / version 11/09

European Paediatric Research Group for HUS and related disorders

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

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: _____

Actual date: _____

6. IDENTIFICATION OF PATIENT

Date of birth: ___/___/___

Hospital: _____

Sex: Female Male

first letter of name:

first letter of surname:

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

Time between first diagnosis and ESRD _____

7. RELAPSES BEFORE TRANSPLANTATION

For Scores see footnotes 1-3 at page 8.

Relapse No	Date	BP score ¹	Proteinuria score ²	Creatinine clearance (Schwartz formula) ³
------------	------	-----------------------	--------------------------------	---

1

2

3

4

5

6

7

8. INVESTIGATION TOWARDS AETIOLOGY

Genetic testing: performed (place: _____) not performed

CFH CFHR1/3 Faktor B C3

MCP Faktor I Thrombomodulin

Results: no mutation

Homozygous deletion of _____

Heterozygous deletion of _____

Polymorphism in _____

CFH antibodies: performed (place: _____) not performed

Result: positive negative

Titer Follow-up (if not measured in Innsbruck):

Date	Titer	Reference Value	Notice

9. 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

10. PRE-TRANSPLANT INVESTIGATION

Date (DD/MM/YY) 19/11/2008

creatinine mg/dl $\mu\text{mol/l}$
Creatinine clearance (Schwartz formula)
hemoglobin level g/dl
thrombocyte count $\times 10^3/\text{mm}^3$
Maximum LDH level units normal value
C3 mg/dl normal values

10.1. HYPERTENSION Score as footnote 1:

10.2. EXTRA RENAL INVOLVEMENT no yes

If yes, give details:

11. PRE-TRANSPLANT TREATMENT

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
<u>Further medication:</u> yes	no		

If yes, please specify:

12. FIRST RENAL TRANSPLANTATION

Date of transplantation: (DD/MM/YY) __/__/____

Time between ESRD and transplantation:		months	years
Time between last episode of HUS and transplantation:		Months	Years
Donor:		Living donor	Cadaveric donor
Native kidneys:		In situ	Nephrectomized
Combined transplantation:	no	yes	

If yes, please specify: _____

12.1. Immunosuppression:

Induction: no yes

If yes, please specify _____

Ciclosporine	no	yes
--------------	----	-----

Steroids	no	yes
Tacrolimus	no	yes
MMF	no	yes
Azathioprin	no	yes
Others	no	yes

Please specify:

12.2. Treatment:

Plasma infusion	no	yes
Plasma exchanges	no	yes
Replacement with albumin	no	yes
Replacement with plasma	no	Yes
Plasma cryosupernatant	no	yes
Immunoglobulins	no	yes
Others	no	Yes

If yes, please specify

13. DISEASE RECURRENCE

please copy if more than 1 recurrence

no yes

If yes, date: (DD/MM/YY) ___/___/___ number: _____

Symptoms:

Minimal hemoglobin level	g/l	
Maximum schistocytes %	%	
Minimum platelet counts	$\times 10^3/\text{mm}^3$	
Minimum haptoglobin level	units	Normal value
Maximum LDH level	units	Normal vaule
Oligo-anuria (<10ml/kg/24h)	no	yes
Maximum serum creatinine (before dialysis)	$\mu\text{mol/l}$	mg/dl

13.1.Hypertension (see appendix 2) no yes

If yes: mild moderate severe

If yes, number of different anti-hypertensive drugs needed for control of BP:

13.1. Neurological complications no yes

If yes, please specify

13.2. Other extra renal involvement

If yes, please specify

13.3. Treatment:

Plasma infusion	no	yes
Plasma exchanges	no	yes
Replacement with albumin	no	yes
Replacement with plasma	no	yes
Plasma cryosupernatant	no	yes

Immunoglobulins

no

yes

Others

no

yes

If yes, please specify

14. OUTCOME AFTER FIRST RENAL TRANSPLANTATION

Date of last follow-up: (DD/MM/YY) __/__/____

Patient alive	no	yes
HUS recurrence	no	Yes
Number of recurrences	0	
If dead, date	___/___/___	Cause
Graft failure	no	yes
If yes,		
due to HUS recurrence	no	yes
due to rejection	no	yes
due to other cause	no	yes

If yes, please specify

Please copy 12-14 if more than one transplantation was performed

NOTES

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 95th percentile

2= Moderate hypertension: 10-30mmHg over 95th percentile

3= Severe hypertension: >30mmHg over 95th 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:

$$\text{Clearance (ml/min/1.73m}^2\text{)} = \frac{\text{Ht (cms)} \times \text{k}}{\text{Creatinine (micromol/l)}}$$

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