ISTITUTO DI RICERCHE FARMACOLOGICHE MARIO NEGRI IRCCS
CENTRO CLINICO *ALDO E CELE DACCO'*
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International Registry of Recurrent and Familial HUS/TTP

REGISTRATION FORM

Please, leave blank !			Please, leave blank !			
CENTER CODE	**FAMILY CODE**		**SUBJECT CODE**	**FILE NO.**	**VISIT NO.**	**DATE of COMPILATION**

Referring physician:

surname | name

hospital address

telephone number | fax number | E-mail

Patient data:

surname | name | sex

birth date | birth place | country | telephone number

address

E-mail

Gruppo sanguigno | *Rh (+/-)*

Codice fiscale *(for Italian resident only)* | Tessera sanitaria *(for Italian resident only)*

History of : HUS ☐ TTP ☐ both HUS and TTP ☐

First episode date of onset: ☐ ongoing: yes ☐ no ☐ date of remission: ☐

Last episode date of onset: ☐ ongoing: yes ☐ no ☐ date of remission: ☐

total number of episodes: ☐

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Clinical data collection (informations regarding the last episode)

Predisposing conditions:

- family history of HUS/TTP — YES ☐ NO ☐
  - if yes, please specify the affected relatives and the disease
- hypocomplementemia — YES ☐ NO ☐
  - family history of hypocomplementemia — YES ☐ NO ☐
  - if yes, please specify the affected relatives
- pregnancy — YES ☐ NO ☐
- birth control pill — YES ☐ NO ☐
- drugs — YES ☐ NO ☐
  - cyclosporin ☐
  - mitomycin C ☐
  - other, specify
- antiphospholipid syndrome — YES ☐ NO ☐
  - notes
- systemic disease — YES ☐ NO ☐
  - if YES please specify
  - malignant hypertension — YES ☐ NO ☐
  - transplant — YES ☐ NO ☐
  - cancer — YES ☐ NO ☐
- other, specify

Triggering agents:

- verotoxin producing E. Coli — YES ☐ NO ☐
- other gastrointestinal infection — YES ☐ NO ☐
- viral infection — YES ☐ NO ☐
- other, specify

Prodromes: YES ☐ NO ☐

if YES please specify

- date of onset
- diarrhea — YES ☐ NO ☐
  - bloody — YES ☐ NO ☐
- vomiting — YES ☐ NO ☐
- abdominal pain — YES ☐ NO ☐
- upper respiratory tract infections — YES ☐ NO ☐
- fever — YES ☐ NO ☐
- other, specify

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Clinical data collection - continue- (informations regarding the last episode)

Clinical manifestations:

- purpura YES ☐ NO ☐
- jaundice YES ☐ NO ☐
- anuria YES ☐ NO ☐

if YES please specify how many days

- need for dialysis YES ☐ NO ☐

if YES please specify how many days

- neurological signs YES ☐ NO ☐

if YES please specify:

- coma YES ☐ NO ☐
- convulsions YES ☐ NO ☐

- other, specify

Complications:

- severe hypertension YES ☐ NO ☐
- sepsis YES ☐ NO ☐

if YES please specify

- bleeding YES ☐ NO ☐

if YES please specify

- neurological sequelae YES ☐ NO ☐

if YES please specify

- other, specify

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Clinical data collection - continue- (informations regarding the last episode)

Treatment:

	YES	NO	if YES please specify commercial nomination of the drug, dose, route and way of administration, duration
antibiotics			
antimotility agents			
antihypertensive agents			
antiplatelet agents			
anticoagulants			
thrombolytic agents			
antioxidants			
steroids			
immunoglobulins			
whole blood transfusion			
packed cell transfusion			
plasma infusion			
plasma exchange			
rituximab			
eculizumab			
ravulizumab			
other			

need for splenectomy YES ☐ NO ☐

note

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Stool colture (informations regarding the last episode)

Done ☐
Not done ☐
Unknown ☐

If Done, please specify

- 1st sample
  Date ☐ Negative ☐ Positive ☐ for ☐
- 2nd sample
  Date ☐ Negative ☐ Positive ☐ for ☐
- 3rd sample
  Date ☐ Negative ☐ Positive ☐ for ☐

Note: ☐

Detection of Shiga-toxin in stools

Done ☐ Date ☐ Result ☐
Not done ☐
Unknown ☐

Notes: ☐

Serum antibodies to Shiga-toxin

Done ☐ Date ☐ Result ☐
Not done ☐
Unknown ☐

Note: ☐

Serum antibodies to E-Coli 0157 lipopolysaccharides (LPS)

Done ☐ Date ☐ Result ☐
Not done ☐
Unknown ☐

Note: ☐

Other specific determinations

☐

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Main clinical and laboratory data at hospital admission and discharge (regarding the last episode)

(if Not Available, please complete with N.A., if Not Done, please complete with N.D.)

	measure unit	hospital admission date:	hospital discharge date:	last follow-up date:	normal range
blood pressure	mm Hg				
hematology					
hemoglobin	g/dl				12-18
hematocrit	%				37-52
RBC	10^6/cc				4.2-6.1
MCV	μ^3				80-99
MCH	pc				27-31
MCHC	g/dl				32-36
WBC	10^3/cc				4.8-10.8
N	%				34.6-71.4
L	%				19.6-52.7
M	%				2.4-11.8
E	%				0-7.8
B	%				0-1.8
platelet count	10^6/cc				150,000-450,000
reticolocytes	‰				1-20
fragmented erythrocytes					
hemoglobin electrophoresis					
Hb A2	%				1.5 - 3.5
fetal Hb	%				< 1 %
abnormal Hb	%				absent
globular osmotic resistance					
serum haptoglobin	mg/dl				
biochemistry					
total protein	g/dl				6-8
albumin	g/dl				5.2-5.5
alfa 1	%				2-5
alfa 2	%				7-10
beta	%				8-15
gamma	%				11.6-19
monoclonal component					absent
glycemia	mg/dl				65-120
urea	mg/dl				15-50
creatinine	mg/dl				0.6-1.3
uric acid	mg/dl				3.4-7
cholesterol	mg/dl				5-200
triglyceride	mg/dl				10-170
total bilirubin	mg/dl				0.6-1.3
direct bilirubin	mg/dl				0-0.3
AST	IU/L				0-46
ALT	IU/L				0-46
CPK	IU/L				M: 25-200; F: 25-170
LDH	IU/L				230 - 460
gamma GT	IU/L				6-50
alkaline phosphatase	IU/L				40-120
Na	mEq/L				136-145
K	mEq/L				3.5-5
Ca	mg/dl				8.7-10.3
P	mg/dl				2.25-4.7

notes:

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Main clinical and laboratory data at hospital admission and discharge (regarding the last episode)

(if Not Available, please complete with N.A., if Not Done, please complete with N.D.)

	measure unit	hospital admission date:	hospital discharge date:	last follow-up date:	normal range
clotting & hemostasis					
prothrombin time	INR				< 1.4
partial thromboplastin time	Ratio				0.82-1.25
thrombin time	Ratio				0.95-1.3
fibrinogen	mg/dl				150-450
lupus anticoagulant					
acid-base balance					
pH (venous)					V: 7.32-7.42; A: 7.35-7.45
PO2	mlHg				80-100
PCO2	mlHg				V: 41-51; A: 35-45
BE	mEq/L				0±2
SB	mEq/L				22-26
BA	mEq/L				V: 24-28; A: 22-26
immunology					
C_3	mg/dl				83-177
C_4	mg/dl				15-45
IgA	mg/dl				80-530
IgM	mg/dl				50-200
IgG	mg/dl				530-1650
RA test					1:40 negative; 1:320 positive
W. ROOSE					1:10 negative; 1:80 positive
LE phenomenon					absent
ANA					negative
ANTI DNA	UI/ml				0-7
ANCA					
sedimentation rate 1h	mm/h				M: 0-15; F: 0-20;
antistreptolysine title					<1:128
PCR					negativo
anticardiolipine Ab	GPL				0-23
urine					
specific weight					1,015-1,028
pH					5.5-6.5
hemoglobin	mg/dl				absent
glucose	mg/dl				0-10
ketones	mg/dl				<200
protein	mg/dl				absent
sediment					
RBC					
WBC					
other					
urea	mg/24h				20-25
Na	mEq/24h				50-230
K	mEq/24h				25-100
creatinine clearance	ml/min				95-150
urineculture					negative
urinary protein	g/24h				0-0.2
urinary protein electrophoresis					

notes:

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Clinical data collection on outcome

	Yes	No		
Peritoneal Dialysis			start at (date)	
Hemodialysis			start at (date)	
Renal Transplant			date	

Treatment

Yes	No

Transplant failure

Reasons for transplant failure

date of return to chronic dialysis

Yes	No		
		other renal transplant	date

Note

Yes	No		
		Liver/Kidney transplant	date

Treatment

Data at last follow-up date

Note

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BIOLOGICAL SAMPLES COLLECTED FOR COORDINATING CENTER:

Please, specify

DATE of COLLECTION: ___/___/___ **DATE of EXPEDITION:** ___/___/___

Patients' treatment at the time of collection:

Date of last treatment with:

- steroids ___/___/___
- immunoglobulins ___/___/___
- whole blood transfusion ___/___/___
- packed cell transfusion ___/___/___
- plasma infusion ___/___/___
- plasma exchange ___/___/___
- plasma cryosupernatant ___/___/___
- rituximab ___/___/___
- eculizumab ___/___/___
- ravulizumab ___/___/___

Laboratory tests at the time of collection:

- Hemoglobin g/dl
- Hematocrit %
- WBC 10^3/cc
- Platelet count 10^6/cc
- Serum creatinine mg/dl
- Serum haptoglobin mg/dl
- LDH IU/L
- C_3 mg/dl
- C_4 mg/dl