

Sd. Hemolitic(o)-uremic - SHU

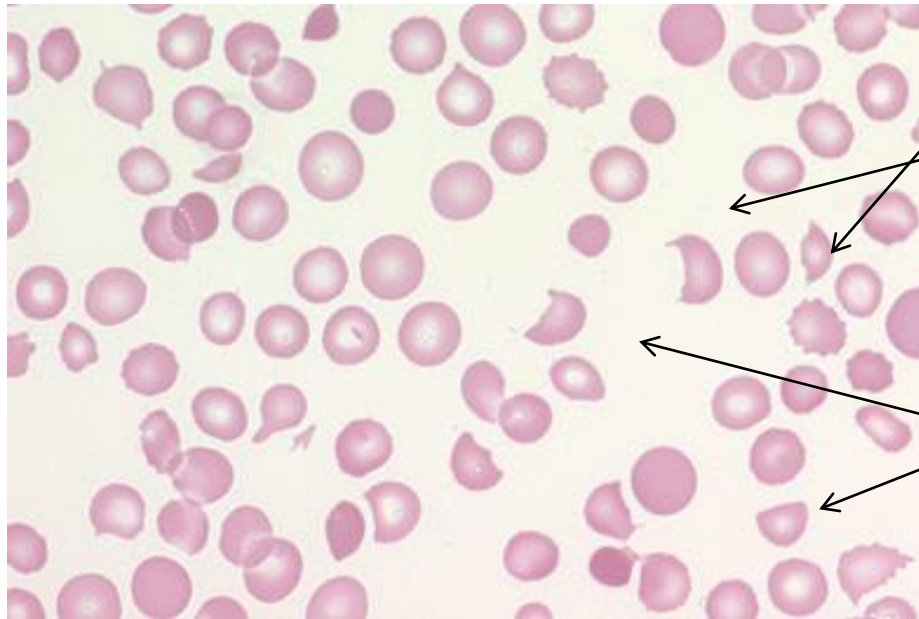
Elemente pentru includere/definire

AKI/IRA de severitate diferită
Anemie microangiopatică
Trombocitopenie

Are multiple etiologii!

Elemente de diagnostic pentru cel tipic D+

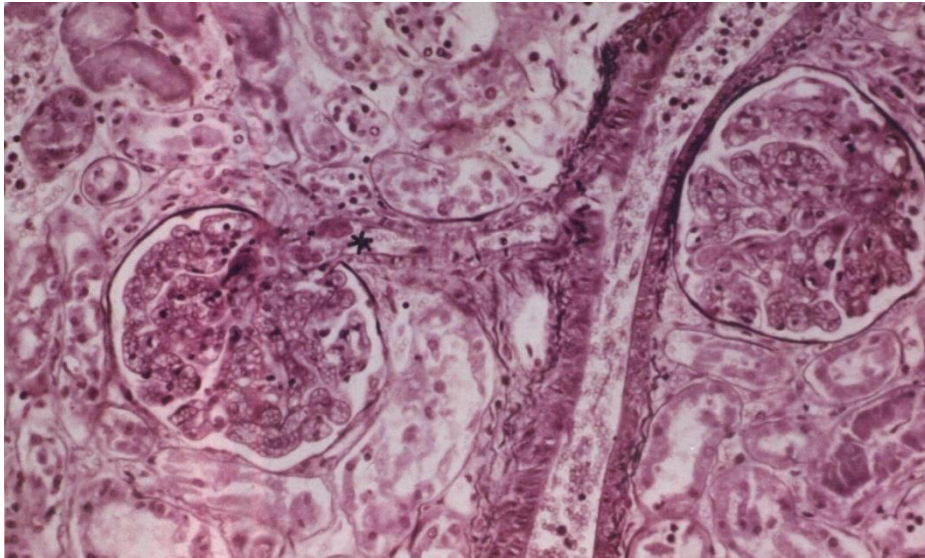
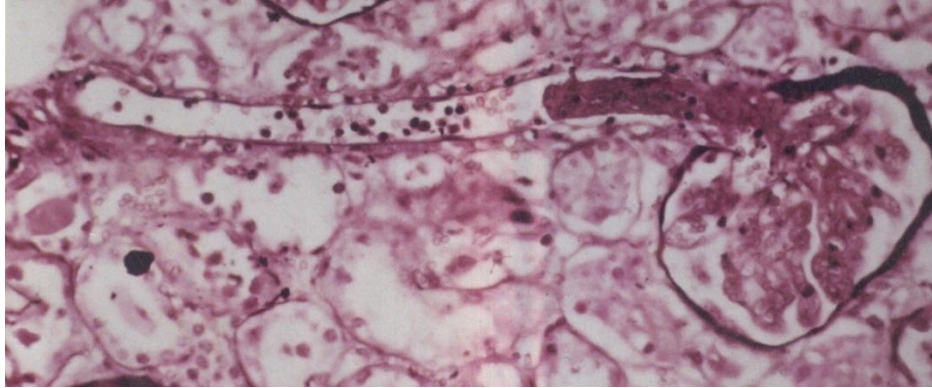
- Diaree, des sagvinolentă
- Hematologic –
 - anemie hemolitică
microangiopatică
 - trombocitopenie



Fragment eritrocitar

Absenta trombocitelor

AKI



D+ HUS/SHU

- EHEC –E Coli enterohemoragic colonizează copitatele (capre, caprioare, etc), carne de vită
- Transmitere – contaminează carnea, laptele, suprafata fructelor sau legumelor prin apă contaminată
- HUS/SHU apare in \approx 15-20% dintre cei infectati cu EHEC
- O157:H7 predomină in UE
 - O26:H11, O103:H2, O111:NM, O121:H19, O145:NM
 - in 2011 outbreak - Romania si alte tari din spatiul European (Polonia, Ucraina, Germania), a fost un serotip particular - O104:H4 !!!

Afectare non-renală

- Convulsii
 - hiponatraemia
 - neurotoxicitate
- Hipertensiune arterială
- Gastro-intestinal
 - Prolaps rectal
 - Megacolon toxic, perforatie, etc
- Cardiomiopatie
- Diabet

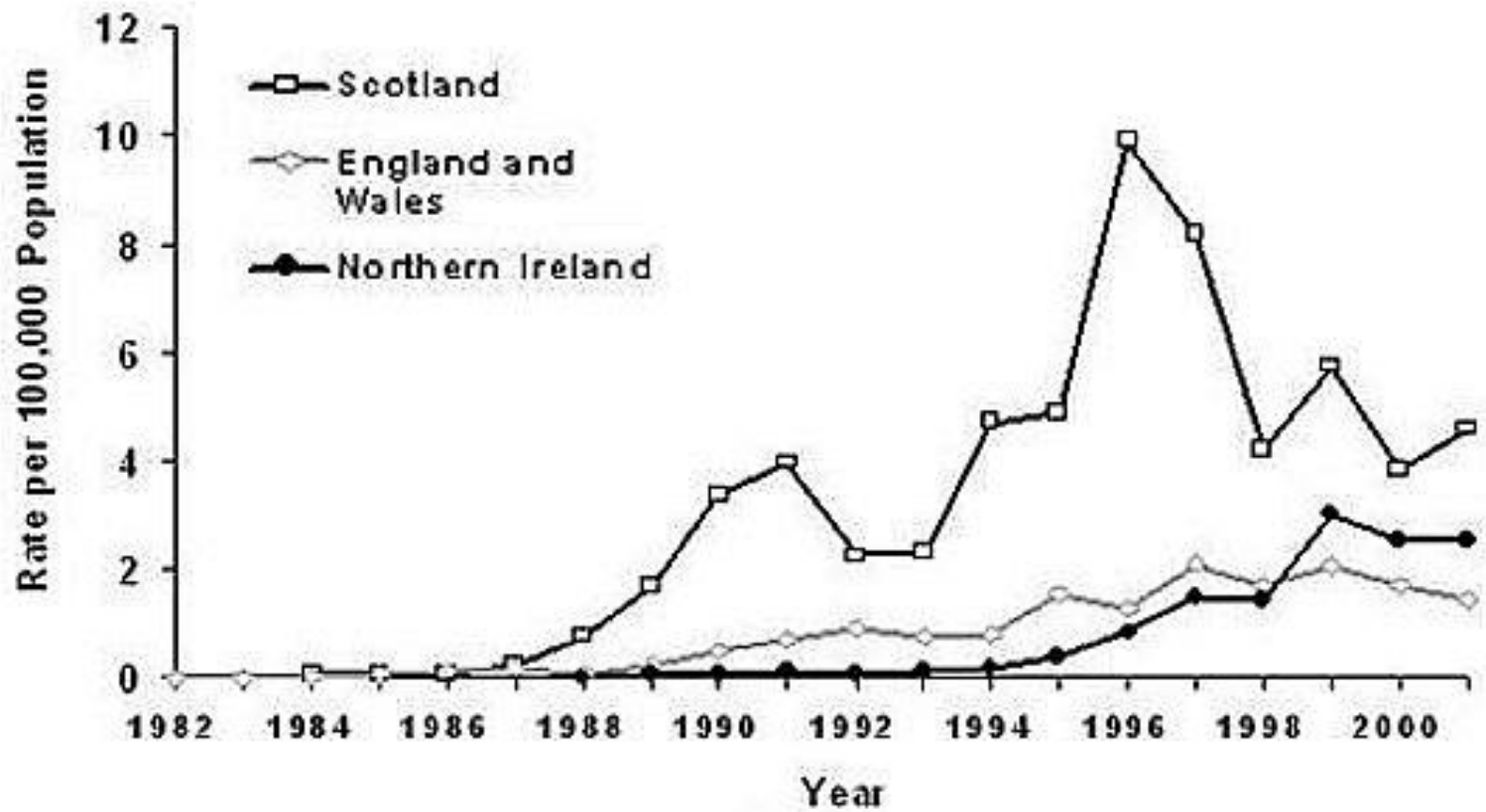
Table 2. Clinical features and acute complications of diarrhea-associated hemolytic uremic syndrome (HUS) in children from the 1997–2001 British Paediatric Surveillance Unit survey compared with children from the 1985–1988 survey

	1985–1988 survey	1997–2001 survey	p value
No. of cases	288	413	
Cases with a diarrheal prodrome	273 (95%)	395 (96%)	0.6
Mean (range) time from onset of diarrhea to diagnosis of HUS	8 days (1–34)	6 days (range 1–35)	<0.001
Severe acute abdominal symptoms	40 (15%)	36 (9%)	0.03
Seizures or other neurologic complications	51 (19%)	52 (13%)	0.06
Hypertension	86 (32%)	92 (23%)	0.02
Cardiomyopathy	4 (1%)	7 (2%)	1.0
Diabetes mellitus	4 (1%)	8 (2%)	0.77

German outbreak - Mai 2011

- Sursa: germeni de fasole si grâu in Saxonia
- Semintele erau aduse din Egipt
- 3793 cazuri – O104:H4
- 827 (22%) HUS, 88% la adult
- 53 decese!!!
- 2010/11 Europa
 - 4000 STEC cazuri in 4 țări, 5.5% cuHUS
 - O157 (41%), O26 (7%), O103 (2.5%)

UK



Level 1: aetiology advanced

1.i Infection induced

(a) Shiga and shiga-like toxin-producing bacteria; enterohaemorrhagic *Escherichia coli*, *Shigella dysenteriae* type 1, *Citrobacter freundii*

(b) ***Streptococcus pneumoniae*, neuraminidase and T-antigen exposure**

1.ii Disorders of complement regulation

(a) **Genetic disorders of complement regulation**

(b) Acquired disorders of complement regulation, e.g. anti-factor H antibody

1.iii **von Willebrand proteinase, ADAMTS13, deficiency**

(a) **Genetic disorders of ADAMTS13**

(b) **Acquired ADAMTS13 deficiency; autoimmune, drug induced**

1.iv Defective cobalamin metabolism

1.v Quinine induced

Level 2: aetiology unknown

2.i Human immunodeficiency virus (HIV)

2.ii Malignancy, cancer chemotherapy and ionising radiation

2.iii Calcineurin inhibitors and transplantation

2.iv Pregnancy, HELLP syndrome and oral contraceptive pill

2.v Systemic lupus erythematosus and antiphospholipid antibody syndrome

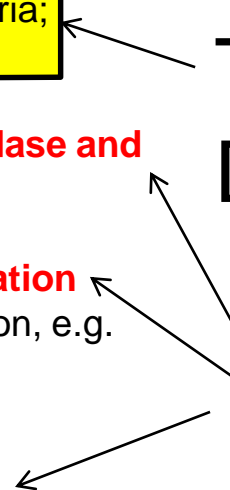
2.vi Glomerulopathy

2.vii Familial, not included in part 1

2.viii Unclassified

Typical/diarrhoeal/
D+ HUS

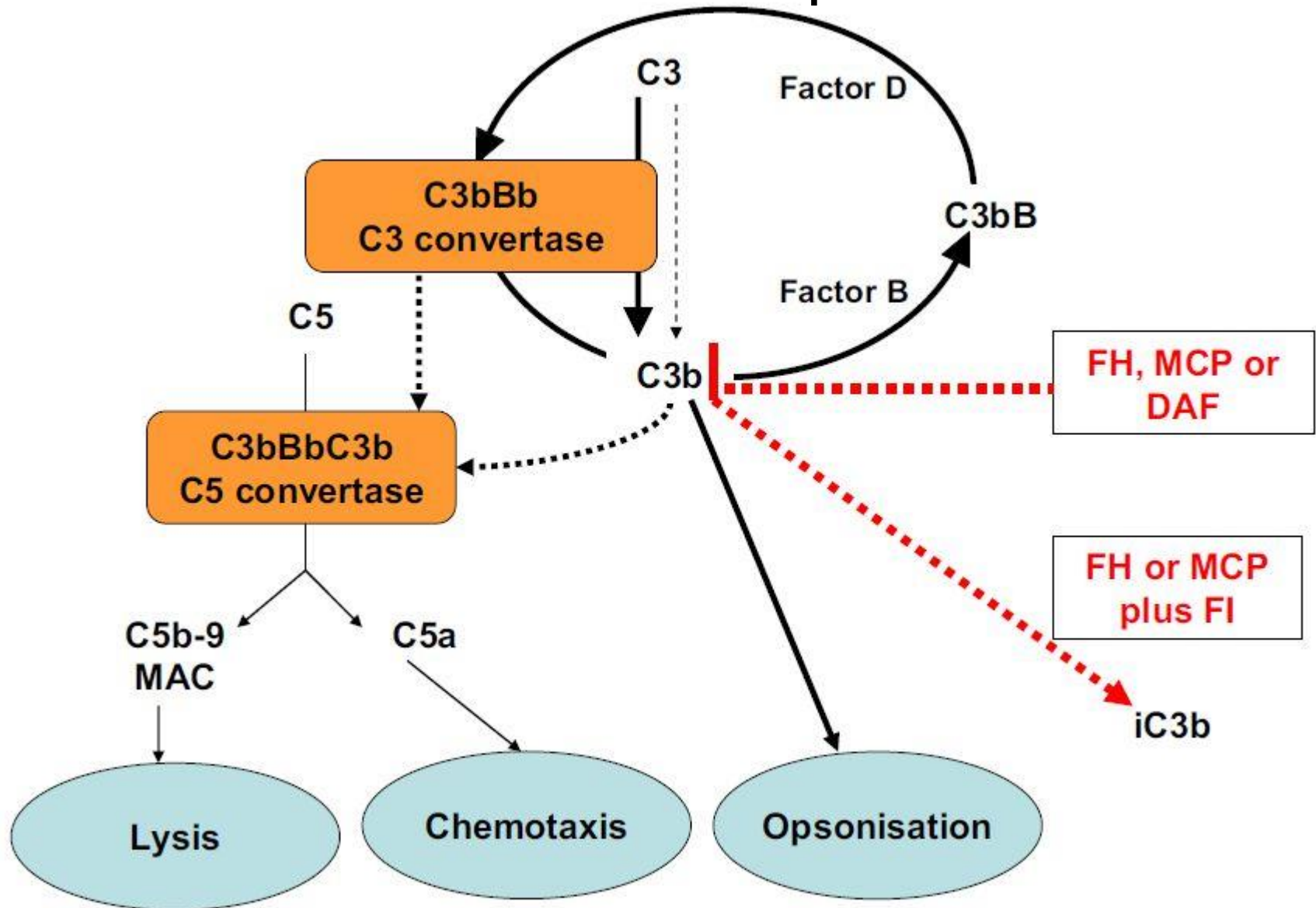
Atypical/
non-diarrhoeal/
D- HUS



D - HUS

- Rapid la nefrolog pediateru!
- Asociază
 - Deces cu rată mare, CKD, hipertensiune, afectare SNC des cu convulsii, recurentă, familială
- Terapie -
 - Plasmafereză
 - Plasmă (după confirmare a ADAMTS13)
 - Eculizumab (leagă C5, blochează C5 convertaza)
 - Transplant – rar!

Calea alternă - complement



Laboratory diagnostic of STEC infection and enteropathic Hemolytic uremic syndrome (HUS)

STEC disease stage	Material	Test	Details	Comments
Diarrhea/colitis				
	Stool or rectal swab	Free Stx or Stx genes	Stx ELISA Vero cell tissue culture assay PCR	Fresh stool preferred for Elisa and vero cell tissue culture assay (decay of the toxin)
		<i>E. coli</i> O157:H7	Sorbitol/tellurite MacConkey agar (or comparable) culture media PCR for virulence genes in isolated colonies	Preserve stool sample and colony sweep or broth at -80 C for specialized laboratory
		Non-O157:H7 STEC strains	Usually sorbitol fermenting in culture PCR for virulence genes in isolated colonies Serological (agglutination) or molecular testing (hybridization, PCR) of bacterial isolates or colonies	
	Serum	Anti-LPS IgM, IgG and IgA	Elisa, immunoblot	Screening for anti-LPS antibodies against the most frequent local STEC-serotypes
	Blood	CBC, smear	Baseline hemoglobin and platelets; presence of schistocytes	
		Creatinine	Baseline renal function	
	Urine	Protein/creatinine ratio Cytology	Baseline/early changes	Microscopic hematuria may be present with colitis or sign of incipient HUS Proteinuria indicates renal involvement
Acute HUS				
	Stool/rectal swab/blood	STEC		See above (unless obtained during the diarrhea phase)
	Blood	CBC, smear	Evolution of hemolytic anemia (schistocytes) and thrombocytopenia as markers of TMA Polynuclear leukocytes $> 20 \times 10^9/L$ marker of TMA severity	May consider coagulation profile
		Biochemistry	Creatinine, electrolytes, albumin LDH, haptoglobin Liver enzymes Amylase, lipase, blood glucose Troponine Optional: CRP (or other acute phase reactant) Optional: C3, C4 (most often normal)	AST and indirect bilirubin elevation generally indicate vigorous hemolysis Troponine elevation indicates myocardial ischemic lesions Detailed complement analysis and/or genetic work-up only if diagnosis of eHUS uncertain and aHUS suspected
		Blood bank	Cross & type	
	Urine		Protein/creatinine ratio, microalbuminuria	During recovery and follow-up: abnormal protein/creatinine ratio or microalbuminuria indicate ongoing TMA or residual kidney disease