HEART DISORDERS IN ATYPICAL HEMOLYTIC UREMIC SYNDROME IN CHILDREN

Kh. Emirova[1], E. Tolstova[1], O. Orlova[1], T. Pankratenko[2], A. Muzurov[3], T. Abaseeva[2],A. Makulova[4], A. Burov[2]

[1]Moscow State Medical Dentist University named after A.I.Evdokimov; [2]Moscow Regional Research Clinical Institute named after M.F.Vladimirsky; [3]Russian Medical Academy of Post-Graduate Education; [4]The Russian National Research Medical University named after N.I. Pirogov

INTRODUCTION

- Cardiovascular manifestations of thrombotic microangiopathy include the development of cardiomyopathy, myocardial infarction, myocardial manifestations, occlusion of coronary vessels, heart failure
- Cardiovascular symptoms occur in 10-43% of patients with aHUS and can cause adverse outcomes

AIM

- To estimate the frequency of cardiovascular complications (CVC) of aHUS in children and outcomes depending on the type of therapy

MATERIALS AND METHODS

- From 2008 to 2015 we examined 33 patients with aHUS from 5 months to 17 years. Genetic screening is conducted in 10 (30%) patients. In 2 cases CFH and CFI mutations were identified, 8 - polymorphic genotypes of complement genes. In 8 (24%) cases aHUS associated with antibodies was diagnosed. 19 (58%) patients received Eculizumab

33 patients with aHUS (n=55)

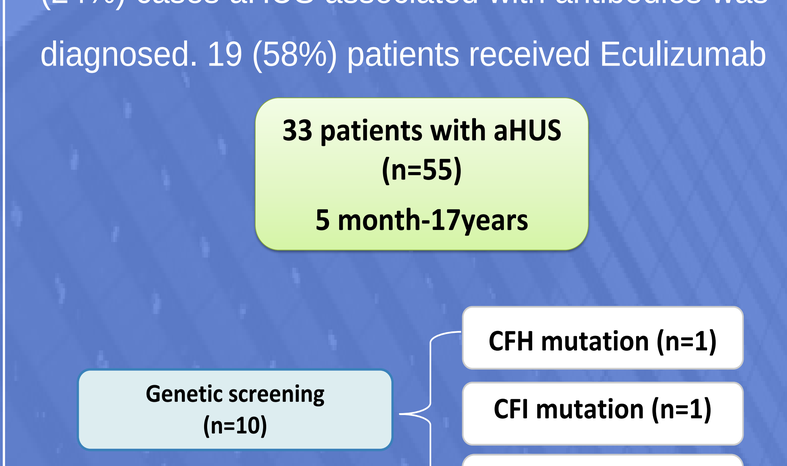
5 month-17years

- **Genetic screening (n=10)**
  - **CFH mutation (n=1)**
  - **CFI mutation (n=1)**
  - **Polimorphic genotypes of complement genes (n=8)**
- **Anti-CFH antibodies** → **N= 8 (24%)**
- **Eculizumab** → **N= 19 (58%)**

RESULTS

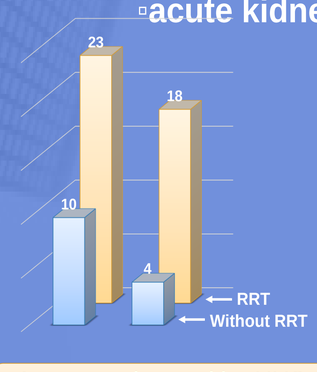
CVC in the acute phase of aHUS were detected in 22 (67%) patients

CVC in the acute phase of aHUS



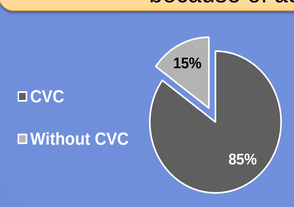
7 (21%) patients received cardiac glycosides for 1-48 months

CVC associated with severity of acute kidney injury (AKI)

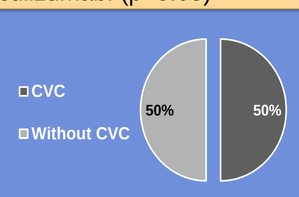


- CVC manifestated in 18 of 23 (78%) children received RRT and 4 of 10 (40%) of the children did not need RRT (p<0.05) had CV

Among patients with aHUS presented in 2012 CVC were detected more often than in the group of children who became ill after 2012, because of access to eculizumab: (p<0.05)

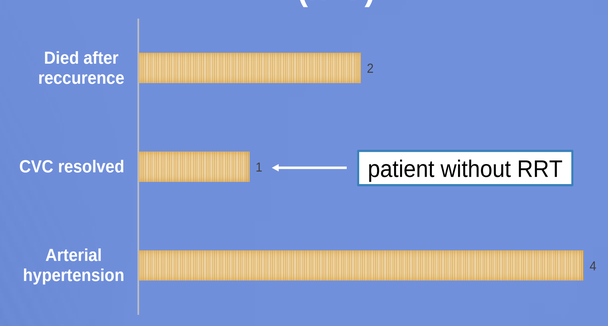


aHUS manifestated before 2012



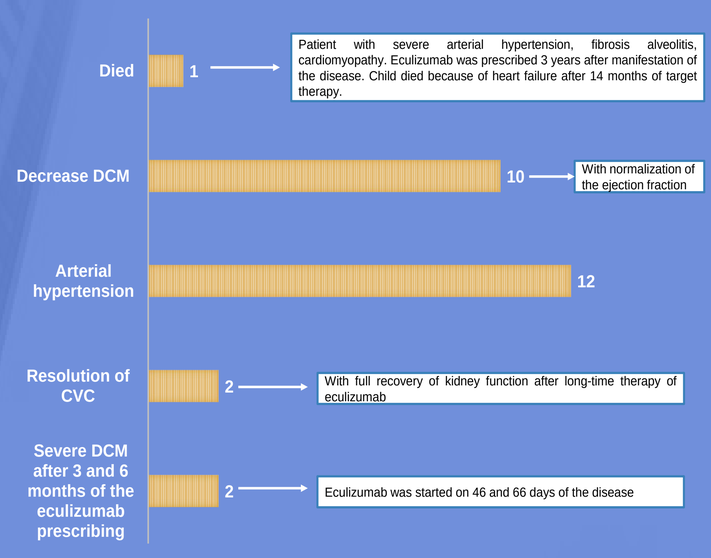
aHUS manifestated after 2012

CVC in patients with aHUS without eculizumab (n=7)

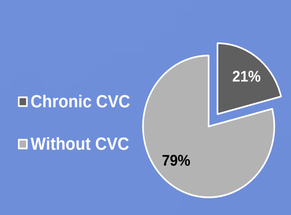


CVC in patients with aHUS with eculizumab (n=15)

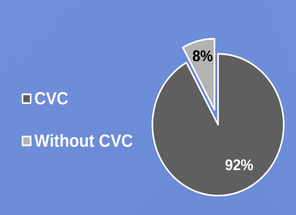
Among 15 children with CVC, eculizumab was assigned for 2 patients in acute aHUS, the other - during the hematologic remission, but persistent organ failure



Chronic CVC was formed in 16 of 31 (52%) patients with aHUS, who survived in the acute period



Normalization of kidney function N=19

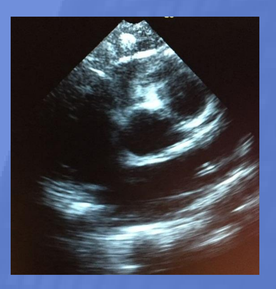


CKD 3-5 stages N=13

▫p<0.05

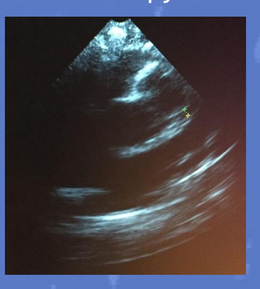
Echographic signs of coronaropathy

▫Before eculizumab therapy



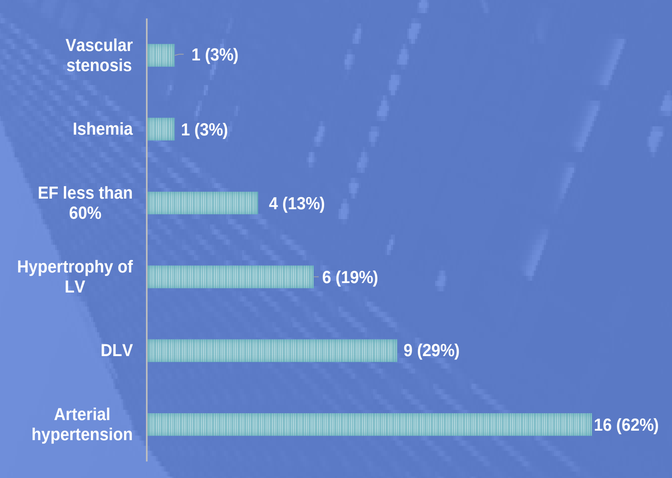
▫Thickening of the walls of the coronary vessels

▫During eculizumab ▫therapy



▫Normalization of echography images

Chronic CVC in patients with aHUS (n=31)



CONCLUSIONS

- CVC is a frequent extrarenal manifestation of aHUS
- Development of CVC in the acute period is associated with a severe AKI, chronic CVC - with the formation of CKD
- High frequency of chronic CVC in children with severe AKI associated with aHUS highlight the need to timely appointment of targeted therapy

REFERENCES

1. Noris, M. & Remuzzi, G. (2014) Cardiovascular complications in atypical haemolytic uraemic syndrome Nat. Rev. Nephrol.
2. Ramon Vilalta et all., Long-term eculizumab improves clinical outcomes in atypical hemolytic uremic syndrome. Pediatr Nephrol (2012) 27:2323–2326
3. Daniel Patschan et al Nephrol Dial Transplant (2006) 21: 1549–1554
4. Marie-Agne` s Dragon-Durey et all., Clinical Features of Anti-Factor H Autoantibody–Associated Hemolytic Uremic Syndrome. J Am Soc Nephrol 21: 2180 –2187, 2010

CONTACT INFORMATION

▫Moscow State University of Medicine and Dentistry
▫Russian Federation
▫Department of Pediatrics
▫Khadizha Emirova
▫PhD
▫Kh.emirova@outlook.com
▫+7(926)6051531

Abbreviation: aHUS – atypical hemolytic uremic syndrome; RRT – renal replacement therapy; CVC – cardiovascular complications; MOFS – multiply organ failure syndrome; CNS – central nervous system involvement; AKI – acute kidney injury; EF – ejection fraction; DCM – dilated cardiomyopathy; LV – left ventricul; DLV – dilated of the left ventricule; CKD – chronic kidney disease

54 ERA
840--MP
Paediatric nephrology
Khadizha Emirova
DOI: 10.3252/pso.eu.54ERA.2017
ePosters supported by F. Hoffmann- La Roche Ltd.