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ATYPICAL HEMOLYTIC UREMIC SYNDROME IN CHILDREN

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Relevance. Thrombotic microangiopathies (TMA) are a class of disorders characterized by microangiopathic hemolytic anemia, non-immune thrombocytopenia, and organ dysfunction. One type of TMA is atypical hemolytic uremic syndrome (aHUS) caused by hyper-activation of the alternative complement pathway due to over activation of C3 convertases and loss of complement regulatory mechanisms. Pathophysiology involves increased spontaneous hydrolysis of C3 to C3b which leads to C3b tissue deposition, membrane attack complex formation and subsequent tissue injury. The underlying susceptibility factors include acquired autoantibodies or germline mutations in complement proteins or their regulators. There are no clear diagnostic criteria for aHUS. Diagnosis involves ruling out other causes of TMA and complement serologic and genetic data. Plasma exchange therapy (PET) has been used to treat aHUS; however, clinical improvement is far less than in thrombotic thrombocytopenic purpura. There is a higher rate of progression to end stage renal disease (ESRD) with almost half of patients progressing. Another option for treatment is eculizumab, a monoclonal antibody that blocks complement C5. Eculizumab has proven effective in aHUS and dramatically changed the prognosis.

Aim: to analyze the cause, clinical presentation, diagnosis, management and outcomes of aHUS in children.

Materials and method. A retrospective analysis of 10 case histories of the patients with aHUS for the period 2013-2023 was carried out in 2nd Children's Hospital Minsk was performed.

Results and its discussion. Among children (n=10) age between 2-7 (Me 4,6) years, including 5 females and 5 males. Clinical presentation: 5 patients present with nausea, vomiting and abdominal pain, 2 with diarrhea, 1 with hemocolitis. Preceding factors: only one patient had upper respiratory tract infection. CBC: 7 patients are presented with severe normocytic anemia and thrombocytopenia, and 3 with moderate to mild normocytic anemia and thrombocytopenia. LDH level was high in 5 patients. Biochemical blood test: 8 patients have high level of creatinine and urea, 2 normal creatinine and urea. Urinalysis: proteinuria and hematuria was severe in 4 cases and moderate to mild in the other cases. Ultrasound of kidney: 7 patients have diffuse changes in the parenchyma of both kidneys, and increase in-the-linear-dimensions-of-both kidneys compared to the age norm and norms for physical development. C3, C4 complement components show normal level in 5. Treatment: 4 patients received eculizumab with immunosuppresses (azathioprine (AZA) + corticosteroids (CS)), 3 PET with same course, 1 only AZA and CS. Kidney transplantation was performed in two patients. Among patients with eculizumab 2 had sepsis then death. Kidney transplantation show favorable prognosis with eculizumab therapy. 4 patients who received PET showed unfavorable prognosis and death.

Conclusion. Half of our patients were above 4 yrs old, only one had preceding upper respiratory tract infection. Half of patients have vomiting and nausea as onset of symptoms, normal C3, C4 complement,7/10 have diffuse changes in the parenchyma of both kidneys, 8/10 ESRD. Disease progression depends on ESRD, delay from symptom onset to initiation therapy of eculizumab and involvement of extra renal system. 6 patients died. 2 patients who received eculizumab in early stage show good prognosis and other 2 patients had sepsis and death, 4 with plasma therapy show unfavorable prognosis, the other 2 with kidney transplantation and eculizumab they survive.