4. BUDD-CHIARI SYNDROME IN CHILDREN-FROM DIAGNOSIS TO LIVER TRANSPLANTATION THROUGH THE PRISM OF A CLINICAL CASE

Author: Samciuc Oleg

Co-author: Gîncu Gheorghe

Scientific adviser: Jana Bernic, PhD, Professor, Natalia Gheorghiu Department of Pediatric Surgery, Orthopedics and Anesthesiology, *Nicolae Testemitanu* State University of Medicine and Pharmacy of the Republic of Moldova.

Introduction. Budd-Chiari syndrome is a pathology in which the obstruction of the hepatic veins takes place, with major transhepatic blood flow disorder, with the subsequent evaluation of the portal hypertension syndrome, cirrhosis, and hepatic insufficiency. Although it is a rare pathology in the child, this congenital malformative disease, even with an adequate treatment does not exclude a reserved prognosis, with a major impact on the child's health.

Case presentation. Patient R.S. 12 years old, presented for the first time in November 2018 in SPNC of pediatric surgery "Natalia Gheorghiu " with main symptoms of general weakness, drowsiness, enlarged abdomen. The patient lost consciousness, which was the reason for consulting the doctor and being hospitalized. Clinical examination reveals an enlarged abdomen with a pronounced vascular pattern, soft palpation, with hepatosplenomegaly, low skin elasticity, pale skin, with multiple vascular stars and petechiae . Signs of free fluid and hepatosplenomegaly were present on the abdominal ultrasound examination. Viral liver markers were negative, biochemical tests indicate a hepatoprive syndrome (proteinemia, increased enzyme activity), hypercoagulation. Liver biopsy shows signs of subacute intrahepatic vascular-intrasinusoidal hypertensive syndrome with dystrophic and inflammatory secondary changes, with sclerotic reactions suggestive of Budd-Chiari syndrome. The undercompensated condition of liver failure was an indication for liver transplantation. For a short period of 14 days the patient was discharged at home with conservative supportive treatment and pre-transplant preparation. Hepatic Transplant was performed after 3 months of diagnosis.

Discussion. The liver transplant followed with an unsatisfactory result, after 2 months postoperatively the death followed. In our case, the patient was diagnosed late at the clinical-evolutionary stage, when the infection was associated with a generalized form of sepsis, followed by post-transplant death, having as causal factors - sepsis, MODS, CID.

Conclusion. Liver transplantation remains the only definitive and effective treatment option in Budd-Chiari syndrome in children, while other treatment methods are useful to maintain normal liver function, especially preoperatively. The sooner the liver transplant is performed, the more favorable your prognosis will be.