DYNAMICS OF DEGREE BREACH STRUCTURE OF LIVER AND DEGREE PORTAL HYPERTENSION IN CHILDREN WITH AUTOIMMUNE HEPATITIS AND WILSON’S DISEASE

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Aim To determine the degree of disruption of the structure of the liver and the severity of portal hypertension and its dynamics during therapy in patients with autoimmune hepatitis (AIH) and Wilson’s disease (WD) in children based on a points system determining the degree of disruption of the structure of the liver.

Methods Analysis of case histories of 176 child at the age from 1 to 17 years (mean age 11.8 ± 3.5 years) with WD (55.4%) and AIH (42.6%). Of them with cirrhosis – 40.4%, without cirrhosis – 59.6%. 49 children (27.8%) liver transplantation was performed.

Results In children with AIH degree of disruption of the liver structure was 20.4 ± 9.2%. In children with liver cirrhosis in the outcome of AIH – 24.8 ± 8.3%, without cirrhosis – 13.2 ± 5.5% (p < 0.001). In children with WD degree of disruption of the liver structure at first hospitalisation was 17.7 ± 12.1% (p = 0.305). In children with liver cirrhosis in the outcome WD – 32.4 ± 9.2%, without cirrhosis – 10.5 ± 4.0% (p < 0.001). In 49 children, who underwent liver transplantation, the degree of disruption of liver structure was 45.2 ± 16.2% (p < 0.001). The degree of disruption of liver structure with AIH after 6 months of therapy – 15.1 ± 8.0% (p = 0.007), in children with liver cirrhosis in the outcome of AIH – 16.9 ± 8.9% (p < 0.001), without cirrhosis – 11.0 ± 2.3% (p = 0.276). After 12 months of therapy, the degree of disruption of the structure of the liver in children with AIH –13.4 ± 5.2% (p < 0.001), in children with liver cirrhosis in the outcome of AIH – 15.6 ± 5.3% (p < 0.001), in children without cirrhosis was 9.8 ± 1.8% (p = 0.139).

Conclusion Scoring system degree of disturbance of the liver structure and severity of portal hypertension can be used as an objective criterion for evaluating the degree of disturbance of liver structure changes in their dynamics on the background of the therapy.

PORTAL CAVERNOMA AND THE CHALLENGES FACED IN EARLY INFANCY

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We describe the case of an ex 25/40 gestation infant initially referred with failure to thrive to her local hospital. She had a background of tracheo-oesophageal fistula and had a degree of chronic lung disease for which she had home oxygen. She was the first born child to unrelated caucasian parents, and was placed in foster care prior to discharge due to parental drug abuse. Examination at referral centre revealed marked splenomegaly. Ultrasound scan revealed portal cavernoma and she was referred to a tertiary unit for investigation and management.

Prior to review, the patient suffered a significant PR bleed at a corrected gestational age of 4 months. Following transfer she suffered 4 further bleeds, 2 of which resulted in cardiac arrest. This is an unusual presentation in a child so young. She had endoscopy and banding, as well as sclerotherapy on 4 separate occasions, each time suffering a further bleed 3 days later. She was commenced on anti-hypertensive medications with minimal effect. Octreotide did control the bleeding but each time this was weaned she suffered further bleeding. She went on to have splenorenal shunt surgery which abated the bleeds temporarily prior to eventual splenectomy. She had a prolonged intensive care admission and was eventually discharged home on.

This case report highlights the sequelae of prematurity as well as management of an unusual presentation of portal cavernoma in a patient of this age.