

Hepatic failure (pediatrics)

Fulminant hepatic failure (FHF) is rare in children. The most common cause is hepatitis A , but often the etiology has not been established.

Definition We define it as an acute liver disease with the onset of encephalopathy within 8 weeks of the clinical manifestation of jaundice , without a previous history of chronic liver disease. In pediatrics we define it as- an acute liver disease that develops a vitamin K-unresponsive coagulopathy from the first symptoms to 8 weeks, where INR is > 1.5 and encephalopathy occurs or INR > 2 without encephalopathy (in addition to standard biochemical parameters of liver damage). ^[1]

Pathogenesis

The causative agent damages hepatocytes → liver cell necrosis → liver failure. Synthetic, excretory and detoxification functions of the liver are impaired → hepatic encephalopathy. High levels of ammonia and glutamate lead to brain edema and circulatory collapse in the CNS. ^[2]

Etiology

The most common are infections and intoxications :

Causes of FHF in newborns

- infections: HSV , echoviruses , adenoviruses , CMV , EBV , HBV ;
- metabolic diseases: galactosemia , tyrosinemia , neonatal hemochromatosis , Niemann-Pick type C disease , mitochondriopathy ;
- ischemia: congenital heart defects , heart surgery, myocarditis , asphyxia .

Causes of FHF in infants and toddlers

- infections: HAV , HBV , HCV , HSV , sepsis ;
- intoxication: paracetamol , amanitin toxin (green toadstool), valproate, isoniazide, halothane;
- metabolic diseases: hereditary fructose intolerance , antitrypsin deficiency , galactosemia .

Causes of FHF in older children

- same as in infants and toddlers, plus Wilson's disease . ^[3]

Clinical picture

The clinical picture is diverse and includes progressive jaundice , anorexia, vomiting, fever, abdominal pain, hepatomegaly , foetor hepaticus , ascites , shrinking liver size due to extensive necrosis. Symptoms of hepatic encephalopathy include: restlessness, irrational hyperreactivity, confusion, lethargy , blurred consciousness, apathy, stupor , coma. Hepatorenal syndrome is renal failure that develops within the FHF, due to changes in the systemic circulation caused by portal hypertension. Multiorgan failure may occur . ^{[4] [5]}

Diagnostics

We observe hyperbilirubinemia , increased aminotransferases (up to 50-100-times, AST / ALT > 1), increased LDH and GGT, then aminotransferases decrease (prognostically unfavorable indicator). Due to impaired synthetic function of the liver, we detect hypoalbuminemia , low cholinesterase activity . We also detect, for example, a coagulation disorder, without a response to vitamin K, rising levels of ammonia and lactate and hypoglycemia. ^{[6] [7]}

Complication

These include ascites , life-threatening bleeding, hypoglycemic coma , renal failure , electrolyte disruption, hypoxia, cardiac decompensation, shock , cerebral edema. ^[8]

Therapy

Liver cell damage is irreversible, treatment is difficult. Supportive measures: decreased protein intake, intestinal decontamination with neomycin and lactulose to reduce ammonia production, electrolyte and fluid substitution, administration of high doses of glucose, correction of acidosis, artificial lung ventilation, administration of vitamin K, coagulation factors and platelet concentrates, antibiotics, hemodialysis. Liver transplantation ("King's College criteria") is also a treatment option. ^[9]

Prognosis

Very serious, without transplantation the mortality is 85% . ^[10]

Links

related articles

- Hepatic failure

Reference

1. LEBL, J, J JANDA a P POHUNEK, et al. Klinická pediatrie. 1. vydání. Galén, 2012. 698 s. s. 370-371. ISBN 978-80-7262-772-1.
2. LEBL, J, J JANDA a P POHUNEK, et al. Klinická pediatrie. 1. vydání. Galén, 2012. 698 s. s. 370-371. ISBN 978-80-7262-772-1.
3. [2]MUNTAU, Ania Carolina. Pediatrics. 4th edition. Prague: Grada, 2009. pp. 399-401. ISBN 978-80-247-2525-3 .
4. [2]MUNTAU, Ania Carolina. Pediatrie. 4. vydání. Praha : Grada, 2009. s. 399-401. ISBN 978-80-247-2525-3.
5. [1]MUNTAU, Ania Carolina. Pediatrie. 4. vydání. Praha : Grada, 2009. s. 399-401. ISBN 978-80-247-2525-3.
6. [1]LEBL, J, J JANDA a P POHUNEK, et al. Klinická pediatrie. 1. vydání. Galén, 2012. 698 s. s. 370-371. ISBN 978-80-7262-772-1.
7. [2]MUNTAU, Ania Carolina. Pediatrie. 4. vydání. Praha : Grada, 2009. s. 399-401. ISBN 978-80-247-2525-3.
8. [2]MUNTAU, Ania Carolina. Pediatrics. 4th edition. Prague: Grada, 2009. pp. 399-401. ISBN 978-80-247-2525-3 .
9. [2]MUNTAU, Ania Carolina. Pediatrie. 4. vydání. Praha : Grada, 2009. s. 399-401. ISBN 978-80-247-2525-3.
10. [2]MUNTAU, Ania Carolina. Pediatrie. 4. vydání. Praha : Grada, 2009. s. 399-401. ISBN 978-80-247-2525-3.