Congenital malformations of the bile ducts

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Annotation: this article is devoted to such a topic as Congenital malformations of the bile ducts

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Atresia and malformations of the intra- and extrahepatic ducts that prevent the normal outflow of bile are relatively common and require urgent surgical intervention. The main manifestation of the defect is obstructive jaundice, which appears in a child at birth and progressively increases. Due to the intrahepatic block, biliary cirrhosis of the liver with portal hypertension develops rapidly, disturbances in protein, carbohydrate, fat metabolism, as well as in the blood coagulation system (hypocoagulation), appear. Treatment. Malformations of the bile ducts that violate the outflow of bile are subject to surgical treatment - the imposition of biliodigestive anastomoses between the extra- or intrahepatic bile ducts and the intestine (jejunum or duodenum) or stomach. With atresia of the intrahepatic bile ducts, surgical intervention is impossible. In these cases, the only chance to save the patient's life is a liver transplant.

Cyst of the common bile duct. The cyst is a local spherical or oval-shaped expansion of the common hepatic or common bile ducts ranging in size from 3-4 to 15-20 cm. The disease is manifested by dull pain in the epigastrium and right hypochondrium, obstructive jaundice due to stagnation of thick bile in the cyst cavity. Diagnosis is difficult, requires the use of modern instrumental methods of research: ultrasound, computed tomography, cholangiography, laparoscopy. Treatment. For the outflow of bile, biliodigestive anastomoses are applied between the cyst and the duodenum or jejunum (with excision of most of the cyst walls or without excision).

Biliary atresia is identified by an increase in both total and direct bilirubin. Serum alpha-1 antitrypsin levels should be measured, as alpha-1 antitrypsin deficiency is another relatively common cause of cholestasis.

Investigations needed to assess liver status include albumin, liver enzymes, prothrombin time/partial thromboplastin time (PT/PTT), and ammonium levels. The concentration of chlorides in sweat secretions should also be determined to rule out cystic fibrosis. Additional testing is often required to evaluate other metabolic, infectious, genetic, and endocrine causes of neonatal cholestasis. Elevated serum levels of alanine aminotransferase (ALT), aspartate aminotransferase (AST), and gamma-glutamyltransferase (GGT) support the diagnosis of biliary atresia and rule out other rare causes of cholestasis.

Abdominal ultrasound is non-invasive and can evaluate the size of the liver and some abnormalities of the gallbladder and common bile duct. Babies with atresia of the bile duct often have a small, shrunken gallbladder or it cannot be seen at all. However, the symptoms are non-specific. A hepatobiliary scan using hydroxy-iminodiacetic acid (HIDA scan) should be done; excretion of a contrast agent into the intestine excludes

biliary atresia, but the absence of excretion may be with bile duct atresia, severe neonatal hepatitis, and other cases of cholestasis. The final diagnosis of biliary atresia is made by liver biopsy and intraoperative cholangiography. The classic histological findings are enlarged portal ducts with fibrosis and bile duct proliferation. Bile plugs can also be seen in the bile ducts. Intraoperative cholangiography reveals the absence of a patent extrahepatic bile duct.

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