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Biliary Hypoplasia

Biliary hypoplasia is a rare cause of persistent neonatal conjugated hyperbilirubinemia. Pathologically, affected children have absent or reduced number of bile ductules with normal distribution of branches of the portal vein and hepatic artery within the liver parenchyma. Biliary hypoplasia is also identified as paucity of interlobular bile ducts (PILBD). Two types of PILBD are recognized: 1) syndromic (arteriohepatic dysplasia or Alagille's syndrome) with characteristic extrahepatic abnormalities (facial appearance, pulmonic artery stenosis, vertebral anomalies, embryotoxon and delayed weight-height development), and 2) non-syndromic biliary hypoplasia. Biliary hypoplasia is clinically indistinguishable from biliary atresia and can sometimes be confused. A definitive diagnosis is difficult to make in early infancy. Differentiation between biliary atresia, hypoplasia and neonatal hepatitis continues to require direct visualization of the biliary ducts. This mean laparoscopic or open intra-operative cholangiography and liver biopsy. The cholangiogram will show diminutive intra- and extra-hepatic biliary tree. Attempts to establish biliary flow by means of hepatic porto-enterostomy (Kasai procedures) in children with PILBD have been unsuccessful and contraindicated. Management is conservative and include predigested formulas, ursodeoxycholic acids (10 mg/kg/day), phenobarbital and A,D,K,E vitamin replacement. Non-syndromic PILBD have better long-term prognosis. Children with syndromic PILBD identified in infancy because of cholestasis have a 50% probability of long-term survival without liver transplantation

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Lumbar Hernias

Congenital lumbar hernias are rare abdominal parietal defects in infants and children. Approximately 10% of all lumbar hernias are congenital and the vast majority are unilateral. They have been divided in three categories: 1) superior - occurring in the superior lumbar triangle (Grynfeltt-Lesshaft), 2) inferior - occurring through the inferior

lumbar triangle (Petit) or 3) a combination of them. They have a well-defined fascial defect. Acquired lumbar hernias outnumber congenital hernias and may result from surgery, infection and/or trauma. Since the hernia defect enlarges with growth or have the potential to incarcerate early operative repair is preferred. Lumbar hernias are associated to the lumbo-costo-vertebral syndrome (caudal regression anomalies, diaphragmatic hernia, ureteropelvic junction obstruction, cloacal exstrophy and lipomeningocele). Repair of small defects can be accomplished by primary closure. Large and recurrent defects may need gortex patching. When they include a more extensive deficiency of the entire lateral abdominal wall extending to the rectus sheath and inguinal ligament closure may also need prosthetic material.

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Castleman's Disease

Castleman's disease (CD) also known as angiofollicular lymph node hyperplasia is a benign disorder characterized by enlarged hyperplastic lymph nodes that occurs very rarely in children. Two classes of CD are identified in children: Hyaline-vascular (HV) type characterized by vascular proliferation and hyalinization, and the plasma cell (PC) type characterized by mature plasma cells in the interfollicular spaces with less vascular proliferation (this type is most common in pediatric age). Its clinical presentation is that of a slowly growing localized abdominal, chest (mediastinum or lung hilum), or neck mass. Within the abdomen CD is located in the small bowel mesentery. May be either asymptomatic or present systemic symptoms of fever, malaise, increase ESR, thrombocytosis, anemia and hypergammaglobulinemia. The enlarged nodes may mimic a malignant tumor of the lymphoid system. Histopathological evaluation confirms the diagnosis. Surgical excision of the mass and surrounding nodes involved is necessary to affect a cure.

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