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Opsoclonus Myoclonus Ataxia

Opsoclonus-myoclonus-ataxia (OMA) syndrome is a rare paraneoplastic or paraviral neurologic syndrome commonly associated with neuroblastoma. Less than 3% of children with neuroblastoma develop OMA syndrome. Clinically the child with OMA syndrome develops an acute onset of rapid chaotic eye movements, myoclonic jerking of the limbs and extremity and ataxia. OMA is believed to be an immune-mediated disorder due to the detection of antineural, antineurofilament and anti-Hu antibodies. The child with OMA syndrome caused by neuroblastoma has a favorable staged disease due to rapid work-up and findings that the patient's immune response to the tumor limits the metastatic and growth potential of the tumor. Rarely do these tumors show n-MYC gene amplification, a poor prognostic finding. Children with neuroblastoma and OMA have an excellent survival. There is no correlation with duration of symptoms and late neurologic outcome. Most children respond to treatment of acute symptoms with steroids or ACTH. Late neurologic sequelae (delay in motor function, speech and cognition) of OMA children can be drastic and affect quality of life. Children with advanced stage disease require more intensive chemotherapy and have better outcomes with regard to late neurological sequelae. The higher the immune response limits the spread of disease but increases the neurologic sequelae.

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Choledocholithiasis

Common bile duct stones (choledocholithiasis) are usually secondary stones from the gallbladder that migrate through the cystic duct causing acute or chronic biliary tract obstruction (obstructive jaundice), pancreatitis or cholangitis. Most common bile duct stones in children pass spontaneously without significant complications. Rarely, children can also develop primary common bile duct stones. Up to 15% of children harboring gallstones can harbor asymptomatic common bile duct stones. Recurrent right upper quadrant pain is the most common clinical presentation. Most children have idiopathic gallstones. Ultrasound is the first study which will reveal whether the child has common bile duct dilatation with the presence of stones. This is accompanied with measurement of liver function tests and pancreatic enzyme. Once dilatation of the common bile duct or stones is suspected, an MRCP is indicated. With good sonographic evidence of common bile duct stones or a positive MRCP an ERCP should be performed to accomplish endoscopic sphincterotomy with extraction of the biliary duct stones. This is followed in the next few days with laparoscopic removal of the gallbladder. The endoscopic approach to choledocholithiasis produces less morbidity, less possibility of long term common bile duct stenosis, and permits preop visualization of the biliary tree. If the endoscopic stone extraction cannot be accomplished with ERCP, then open or laparoscopic common bile duct exploration is indicated.

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Cholelithiasis in SCD

Cholelithiasis is a frequent complication of Sickle Cell Disease (SCD) in children. Prevalence of developing pigmented stones can range from thirty to 50%. Even the prevalence of developing common bile duct stones in children with SCD is higher than the general population. Complications related to cholelithiasis include biliary colic, pancreatitis, cholecystitis and obstructive jaundice. Children with SCD should be screened for

cholelithiasis since the age of eight years using abdominal ultrasound. Once cholelithiasis is identified removal of the sick gallbladder should be accomplished regardless if the child is asymptomatic or not. Preoperative transfusion reduces the morbidity of the surgical procedure. Elective laparoscopic cholecystectomy should be the gold standard in children with SCD and asymptomatic cholelithiasis to prevent the potential complications of biliary colic, acute cholecystitis, and choledocholithiasis, which lead to major risks, discomfort, and longer hospital stay. Laparoscopic cholecystectomy results in a shorter hospital stay with fewer postoperative complications than open operation in patients with sickle cell disease and is the procedure of choice in the treatment of cholelithiasis in such patients.

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