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Asplenia

The absence of the spleen (asplenia) occurs after surgical removal, following chronic conditions or congenital. Trauma is the most common cause of removing the spleen in children and sickle cell disease is the most common cause of functional asplenia in children. Congenital absence of the spleen is usually associated with serious malformations, primarily cardiovascular and abdominal heterotaxia. The spleen contributes importantly to the normal and pathologic removal of blood cells from the circulation and to defense against infection with encapsulated bacteria. Asplenia increases the risk of fulminant bacteremia (post-splenectomy sepsis) and mortality with these organisms. This risk is also increased by the underlying condition that caused the removal of the spleen, i.e., trauma, malignancy or hematologic disease. Several recommendations have been given when dealing with an asplenic individual. These are to vaccinate the child against pneumococcus (Pneumovax vaccine), hemophilus influenza type b and meningococcus. Regarding Pneumovax use revaccination after 3-5 years is recommended for children with asplenia who are 10 years of age or younger and for older children and adults who were immunized at least five years before. Duration of vaccine-induced antibodies is unknown but may be shorter than that in normal persons. Long-term antimicrobial prophylaxis is also used. This carries the problem of compliance and for how long. Significant febrile episodes should be managed aggressively, and probably most important, the patient and family should be carefully educated about this complication (name tag). Most deaths from hyposplenia-related septicemia are preventable.

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Pancreas Divisum

Pancreas divisum (PD), believed the most common congenital anomaly of the pancreas, is an embryologic variation of pancreas development where the dorsal (Santorini) and ventral portions (Wirsung) ducts drain separately. Diagnosis is made with ERCP (short duct of Wirsung that does not communicate with main pancreatic duct of Santorini). Not everybody with this ductal anomaly develops pancreatitis. Likewise with the minor papilla draining the bulk of the pancreas in PD, a small orifice size (< 0.75 mm) plays a role in

outflow obstruction and development of pancreatitis. Children with PD and recurrent episodes of pancreatitis will need endoscopic sphincterotomy of the minor and sometimes major papilla. If not feasible technically, surgical sphincteroplasty of both papillae along with cholecystectomy (bile stasis leads to gallstones) is indicated. Intraoperative pancreatogram will help determine if both papilla are stenotic. Once chronic pancreatitis is established, ductal drainage or resection may be necessary.

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Meconium Peritonitis

Meconium peritonitis (MP) is a chemical peritonitis that occurs following bowel perforation during fetal life. It is generally looked upon as benign, resulting in no long-term sequelae. The peritonitis occurs when the meconium leaves the bowel, enters the peritoneal cavity and spreads throughout causing a sterile inflammatory reaction. Most common site of bowel perforation is the distal ileum, and 50% of babies with MP develop intestinal obstruction. Prenatal ultrasound findings include ascites, intraabdominal masses, bowel dilatation and the development of intraabdominal calcifications. Bowel disorders which lead to MP in utero are those resulting in bowel obstruction and perforation, such as small bowel atresias, volvulus and meconium ileus. MP can be divided into simple or complex. Cases with spontaneously healed perforation (simple MP) need observation as they rarely develop symptoms. Newborns with complex MP are born with bowel obstruction a/or pseudocyst formation (localized collection of meconium contained in a cyst made of fibrous granulation tissue). Complex MP needs surgical therapy.

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