

PEDIATRIC SURGERY Update © Vol 20 No 04 APRIL 2003

Partial Splenectomy

Partial splenectomy (PS) is a safe and effective alternative for several hemolytic disorders and hemoglobinopathies in children including hereditary spherocytosis, sickle cell disease and b-thalassemia. It is the operation of choice for all benign splenic conditions. The main objective of partial splenectomy is preserve splenic contribution to host defense and reduce the incidence of post splenectomy sepsis (4% risk; mortality rate 1.5%), while reducing symptoms of hemolysis and sequestration from the systemic disorder. The risk of septicemia is even higher in very young children (< four years old). Technically, the goal of the procedure is to remove 80% of splenic tissue by preserving either the upper or lower pole. The preserved remnant has not proved to reduce the possibility of post splenectomy sepsis due to the prohibited large clinical trial needed, but has been found to preserve partial phagocytic function and normal immunoglobulin level, reduce Howell-Jolly bodies with confirmed visualization on scintigraphy. In hereditary spherocytosis partial splenectomy has achieved two aims: decrease red cell destruction and preserve the phagocytic and immunologic functions of the spleen. Because of chronic hemolysis the size of the spleen remnant has a tendency to increase with time, specially rapid the younger the child. Follow-up is closely needed in these children to determine if they need conversion to total splenectomy. For sickle cell anemia partial splenectomy has greatly reduced the acute sequestration crisis, need for hospitalization and subsequent transfusions but does not always preserve splenic function. This is probably cause by progressive auto-infarction of the splenic remnant. Since sickle cell patients autoinfarct between 36 and 60 months of life the procedure would benefit more children with less than 48 months of age.

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Congenital Rectal Stenosis

Congenital rectal stenosis (CRS) is a rare anorectal malformation characterized by a tubular defect in which an stenotic distal rectum partially communicates with the distal anal canal. The stenosis is located at the natural limit of rectum and anal canal (immediately above the pectinate line), runs for one to two centimeters in distance while the child is born with a normal-looking external anus. The defect has all the necessary elements responsible for bowel control including excellent muscle component, an anal canal located within the limits of the external sphincters with all the nerve ending that afford normal sensation and a normal sacrum. CRS is usually detected in the newborn during the initial physical examination. Rectal stimulation can cause propulsive watery output. Local ischemia occurring late during fetal life may be the mechanism responsible for the creation of rectal stenosis of the middle and/or upper rectum. CRS is the hallmark finding in the Currarino's triad (sacral bony abnormality, presacral mass and rectal stenosis). Management of CRS consists of serial daily dilatations increasing the size of the dilator progressively or continuous balloon distension. Failure of conservative therapy (dilatation) should alert the physician to the presence of an associated pathologic condition in the presacral space. Chronic constipation is a common post-dilatation feature in these patients.

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Omental Infarction

Omental infarction is a very rare painful condition that can mimic appendicitis, pyelitis or cholecystitis in children. This condition is different from omental torsion which is usually associated with intraabdominal pathology such as omental cysts, hernias, tumors or adhesions. Etiology of omental infarction is unknown. Torsion occurs with omental long axis rotation resulting in venous obstruction followed by arterial obstruction, infarction and gangrene. The omentum usually rotates around the distal right gastro-epiploic artery causing right lower abdominal pain. Obesity is a well-known predisposing factor. All patients present with acute onset of right lower quadrant pain. Clinically, the child develops local tenderness with peritoneal signs but without gastrointestinal symptoms. CT scan (defined area of fat interspersed with hyper attenuating streaks) is diagnostic. Laparoscopy

will confirm the diagnosis. Surgical resection of the infarcted omentum results in immediate resolution of pain with minimal morbidity. Others believe that with preoperative diagnosis resection should depend on symptoms.

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