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Duodenal Web

Intrinsic congenital obstruction of the duodenum is either caused by an atresia, stenosis or web with a prevalence of 1:6000 live births. Congenital duodenal obstruction is the result of several embryologic defects in foregut development, canalization or rotation. Duodenal atresia causes a complete obstruction producing symptoms after birth with evident radiographic findings, while true stenosis and webs of the duodenum can have delayed presentation manifesting later in life. The radiographic signs of duodenal atresia are the “double bubble” with gaseous distension of the stomach and proximal duodenum and total absence of intestinal gas distally. Duodenal webs are the most rare and difficult to diagnose of the intrinsic obstruction occurring in this portion of the gastrointestinal tract. They manifest with recurring and progressive bouts of vomiting, nausea, epigastric discomfort and early satiety. Plain film shows a double bubble effect with distal air. The differential diagnosis includes duodenal stenosis, malrotation with midgut volvulus, pyloric stenosis and gastroesophageal reflux. The diagnosis of a duodenal web can be made with fiberoptic upper endoscopy, hypotonic duodenography or an upper gastrointestinal contrast imaging study. Initial management of any form of intestinal obstruction must include gastric decompression, fluid resuscitation and correction of electrolyte abnormalities, most commonly hypochloremic metabolic alkalosis. There are several varieties of webs: complete duodenal atresias or imperforate webs, intraluminal imperforate webs (wind sock webs), and perforated webs with either central or eccentric apertures. Most duodenal webs are preampullary with a central single aperture and size ranging from 0.5 to 20 mm. Annular pancreas is the most common associated condition of a duodenal web. Surgical management of a duodenal web includes transduodenal web excision with transverse duodenoplasty or bypass duodenoduodenostomy. Complications of the procedure include delayed gastric emptying, pancreatitis, wound infection and duodenal stenosis or leak.

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Familial Hirschsprung’s Disease

A small proportion, probably in the range between 2% and 9% of children born with Hirschsprung’s disease (HD), have an inherited familial predisposition, as well as occurring in mono- and dizygotic twins, and a 12% association with chromosomal anomalies. This subgroup of children with familial HD is usually male with an apparent incomplete gene penetrance and variable phenotype. Affected families with HD have 200 times higher risk of recurrence. The length of the aganglionic segment is one of the most consistent predictors of HD transmissibility. Long segment and total colonic aganglionosis is significantly more frequent in familial HD cases than in the sporadic variety carrying the highest risk of recurrence. There is an increase female tendency to transmit the condition to the offspring. The highest recurrence rate occurs in a male sibling of a female proband with long segment HD. RET gene is the major gene causing Hirschsprung's disease (HD). RET proto-oncogene and EDNRB gene variation are identified in 70% of familial HD compared with 30% in the sporadic form. No specific sites on the gene are consistently identified with variations including mutations, frame shifts, deletions, and single nucleotide polymorphism in the RET gene. The combined cumulative effects of the susceptibility loci of RET and EDNRB genes, probably contribute to long-segment and total colonic aganglionosis in familial cases. RET intronic variations may influence gene penetration. Genetic counseling should be offered in these families and in particular for those patients with long segment and total colonic aganglionosis.

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Sengstaken-Blakemore Tube

In the 1946, Blakemore and Sengstaken developed a tube with two balloons to control bleeding from esophageal or gastric varices. The tube allows independent inflation of the gastric and esophageal balloon with a distal lumen to aspirate the stomach. With the routine use of modern endoscopic technique the Sengstaken-Blakemore tube (SBT) is rarely use today. The tube is passed down into the esophagus and the gastric balloon is inflated inside the stomach. Traction is applied to the tube so that the gastric balloon will compress the gastroesophageal junction and reduce the blood flow to esophageal or gastric varices. If the use of traction alone cannot stop the bleeding, the esophageal

balloon is also inflated to help stop the bleeding. The esophageal balloon should not remain inflated for more than six hours to avoid necrosis. Cooling the SBT has been held to stiffen it and aid during insertion. In a few children bleeding is so severe that medical (octreotide, vasopressin) and endoscopic (sclerotherapy and/or banding) interventions are not feasible, safe or effective and the SBT is required to control blood loss and allow the patient to be resuscitated and stabilized. The SBT can be used effectively in children with bleeding esophagogastric varices from portal hypertension and in the rare case of an aortoesophageal fistula. Hemodynamically unstable patients requiring intubation ventilator support and large volumes of fluids and blood products for catastrophic upper GI bleeding considered unsuitable for first line management should undergo SBT insertion. Almost half of those patients are infants needing a 14 or 16 Fr SBT. In cases of bleeding aortoesophageal fistula the SBT can be lifesaving providing suitable time for surgical repair. Complications of SBT insertion include: recurrent bleeding, tube dislodgement, esophageal rupture, airway compromise, external cardiac compression, aspiration pneumonitis, gastric mucosal ulceration and pressure necrosis at lips and cheeks. Pressure necrosis occurs after more than four days of constant use of the SBT.

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