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Bile Duct Bifurcation

Duplication, or more appropriately phrase bifurcation of the bile ducts is an anomaly associated to duodenal atresia. This anomaly can lead to find gas in the distal bowel of a duodenal atresia. The bifurcated bile duct communicates between the proximal and distal atetric segments bypassing gas between them. A bifid biliary system inserts at blind upper and lower pouches of the duodenum, and the common bile duct inserts in a Y fashion. The incidence of duodenal atresia with an anomalous bifurcated bile duct conduit is higher than is thought and occurs more frequently than that associated with duodenal stenosis. Down syndrome is highly associated with cases of an anomalous bifurcated bile duct conduit. Contrast studies are generally not performed in the typical clinical and radiographic evaluation of duodenal atresia; however, an upper gastrointestinal series can demonstrate the anomalous bile duct bifurcation or duplication. The presence of such anomaly can predispose the patient to cholestasis and cholangitis due to duodeno-biliary reflux. The management of duodenal atresia consists of duodenoduodenostomy. Care should be taken at surgery to avoid obstruction or injury to the anomalous bifurcated bile duct, and operative cholangiography may be useful to document continued bile duct patency following repair of the atresia.

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Microvillous Inclusion Disease

Microvillous Inclusion Disease (MVID) or microvillous atrophy is a very rare congenital disorder of the intestinal epithelial cells that present with persistent life-threatening watery diarrhea and is characterized by morphological enterocyte abnormalities. Onset of diarrhea most often occurs within the first days of life (early) or in the first two months

of life (late). MVID is very rare and transmitted as an autosomal recessive trait. Patients can lose up to 30% of body weight. Small bowel biopsy is diagnostic revealing villous atrophy without significant crypt hyperplasia. Periodic acid-Schiff positive granules accumulate in the apical cytoplasm of immature intestinal epithelial cells. Children with MVID are totally dependent on parenteral nutrition. Long-term outcome is generally poor, due to metabolic decompensation, repeated states of dehydration, infectious and liver complications related to the parenteral nutrition. Relentlessly the child develops intestinal failure secondary to the diarrhea. There is no specific treatment for MVID other than hydration and parenteral nutrition. Isolated intestinal transplantation or combined liver–small bowel transplantation is a last treatment option when significant liver disease exists. Replacement of the conventional soybean oil–based lipid emulsions with an omega-3–rich product have been found to rapidly reverse parenteral nutrition-induced cholestasis in patients with MVID.

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Endoluminal Intestinal Lengthening

Endoluminal intestinal lengthening is an option still in experimental phase for intestinal lengthening in cases of short bowel syndrome. Mechanical force is a viable method for increasing intestinal length while preserving the intestinal function. Distraction enterogenesis or the application of forces to the small bowel has been shown to increase length through the induction of cellular proliferation. Some designs previously used for such purposes consist of screws, hydraulic pistons and remotely controlled ratcheting devices. Experimental studies have demonstrated the feasibility of using a polymer-coated spring capsule intraluminally placed in a piece of intestine of experimental animals for timed deployment of an expanding device for bowel Lengthening. The restored jejunal segment had an increase in crypt depth and no difference in villus height compared with normal jejunum. Sucrase activity in the restored segment was not different from that in normal jejunum. Using these methods the small bowel can be lengthened three to fourfold times its original length. Mechanical lengthening may be a useful technique to increase intestinal length in patients with short bowel syndrome.

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