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Hirschsprung's Disease

Hirschsprung's disease (HD) is the congenital absence of parasympathetic innervation of the distal intestine. Occurs one in 1000-1500 live births with a 4:1 male predominance; 96% are term and 4% prematures babies. Symptoms usually begin at birth with delayed passage of meconium. In some infants, the presentation is that of complete intestinal obstruction. Others have few symptoms until several weeks of age, when the classic symptom of constipation has its onset. Initial evaluation includes an unprepped barium enema (the first enema should be a barium enema!). The aganglionic rectum appears of normal caliber or spastic, there is a transition zone and then dilated colon proximal to the aganglionic segment. Rectal suction biopsy is then performed and the submucosal plexus is examined for ganglion cells. Difficulty in interpreting the specimen would require a full-thickness biopsy for definitive diagnosis. Conventional treatment requires performing a "leveling" colostomy in the most distal colon with ganglion cells present. Placement of the colostomy in an area of aganglionosis will lead to persistent obstruction. Once the child has reached an adequate size and age a formal pull-through procedure is done. Current preference is for Soave procedure (modified endorectal pull-through). A tendency toward primary pull-through without colostomy in early infancy is being reported along with a laparoscopic version of this procedure with a decrease in morbidity and hospital stay.

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Asymptomatic Malrotation

Contrast studies often done for other reasons may disclose the presence of a rotational anomaly of the bowel in an asymptomatic child. This trigger the question whether surgery is needed to reduce the risk of volvulus (midgut infarction) in the life expectancy of the affected patient. To make a complete assessment of the rotational anomaly and know the location and existence of the duodeno-jejunal (Treitz) and ileo-cecal junctions an UGIS with follow-through and barium enema will be neccesary. If the rotational anomaly shows that these two junctions are near each other (narrowing of the mesenteric base) and proximal

to the superior mesenteric artery the threat of volvulus becomes real and prophylactic Ladd's procedure should be offered. Ladd's procedure can be done laparoscopically. Patients with malrotation more likely have bands, mesenteric defects, foreshortened dorsal root and redundant leafs. Doppler color US or CT can tell whether there is an anatomic change in the position of the superior mesenteric vein that suggests volvulus. Children with non-rotation associated to a surgical condition (diaphragmatic hernia, abdominal wall defects, prune belly, etc.) benefits from the adhesions created during primary repair and seldom develop volvulus. The morbidity after a Ladds procedure might be significant in some patients.

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Heterotaxia Syndrome

Helwig is credited with describing the Heterotaxia (Polysplenia) syndrome in 1929. The syndrome a defect of lateralization commonly thought of as "bilateral left sidedness" is highly variable in its anatomic expression. Is a rare congenital disorder characterized by abnormal viscero-vascular situs with either left or right isomerism that usually coincides with complex cardiac malformation. Consist of polysplenia, intestinal malrotation, absence of the inferior vena cava, situs inversus, preduodenal portal vein, abnormalities of the hepatic artery and cardiac defects. The syndrome frequently involves visceral disorders such as malposition, malrotation or malfixation of abdominal organs. Malrotation is the most frequent anomaly encountered and classically presents with duodenal obstruction during early infancy. Malfixation of the stomach might produce gastric volvulus. Management in this cases consist of both Ladd's procedure and gastropexy. The polysplenia syndrome is the most common extrahepatic anomaly found in association with Biliary Atresia. In no way the syndrome jeopardized the result of porto enterostomy in these children.

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* Edited by: **Humberto L. Lugo-Vicente, MD, FACS, FAAP**
P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico 00922-0426.
Tel (787)-786-3495 Fax (787)-720-6103
E-mail: titolugo@coqui.net Internet: http://home.coqui.net/titolugo
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