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TCA

Total colonic aganglionosis (TCA) is a variety of what is commonly known as long segment Hirschsprung's disease plagued with delay in diagnosis, higher morbidity/mortality and controversial management. The colon is aganglionic sometimes involving a variable segment of distal ileum. Clinically, infants presents with small bowel obstruction, persistent obstipation, distention, or poor weight gain. Early diagnosis depends on clinical awareness of the condition in neonates with intestinal obstruction, diarrhea, or both. Barium enema changes may easily be passed as unremarkable. Radiographic findings of a shortened colon of normal caliber or the presence of "jejunalization" of the colon suggest TCA in patients with a suggestive history. However, free ileal reflux during the examination with a transition point in the ileum and retention of barium in the entire colon after the examination may be diagnostic. Diagnosis is confirmed by rectal biopsy, and the extent of aganglionosis documented after multiple intestinal biopsies. Management consists of initial enterostomy in the proximally ganglionic bowel (a source of fluid/electrolytes losses, prolonged hospitalization, and nutritional deficiency), followed by a pull-through (Soave or Duhamel) using a side-to-side anastomosis between a segment of the aganglionic colon to the ganglionated small bowel. The procedure provides a greater surface area for absorption and fecal storage. Diarrhea and distension are temporary after the pull-through, but most patients ultimately tolerate normal feedings.

Fibrolamellar Carcinoma

Fibrolamellar carcinoma (FLC) is a rare histologic subtype of hepatocellular carcinoma associated with long-term survival and encapsulation (few mm to 1 cm thick capsule). Occurs almost exclusively in young adolescent women in the absence of cirrhosis, and the serum alpha-fetoprotein is usually normal. Some thinks that it arises in areas of focal nodular hyperplasia with variable malignant potential. Distinctive histologic features include deeply eosinophilic polygonal hepatocytes and abundant fibrous stroma. Aromatase level are high causing gynecomastia and failure to enter puberty. A defect in chromosome one has been recently identified. Computed tomography remains the most

accurate technique for diagnosis and staging (large, low attenuation, well-defined edges, some contain areas of calcification or necrosis). The treatment of choice remains radical operation a goal that can be achieved by including partial and total hepatectomy depending on the stage of the tumor. Statistically significant better five year survival rates are observed in patients with solitary tumors and absent regional lymph node metastases. Cause of death generally is due to a lack of control of the primary tumor. Successful treatment appears to relate to the ability to do a total excision of the primary hepatic tumor.

Byler's Disease

Byler's disease is a progressive familial intrahepatic cholestasis liver condition (autosomal recessive) initially seen in infants less than six months of age. Clinically includes jaundice, itching, failure to thrive, and death from liver cirrhosis. Laboratory findings are: elevated levels of bilirubin and aminotransferases, with normal to low levels of GGT and cholesterol. Liver biopsy is non-specific: cholestasis, liver cell plates and ballooning of hepatocytes in central zones. Presumably, the condition could arise from a transport defect that causes retention of bile salts resulting in secondary toxic hepatocyte injury. Some patients respond to medical therapy (UCDA); in other children liver transplant remains standard therapy. Recently, the use of partial bile drainage using cholecystostomy with an interposed jejunal segment to the skin is of advantage in reducing the enterohepatic circulation of bile acids interrupting the natural history of the disease, reducing its complications and preventing the need for early transplantation in many patients.

*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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