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Meconium Ileus

Meconium Ileus (MI) is a neonatal intraluminal intestinal obstruction associated with Cystic Fibrosis (10-20%). The distal ileum is packed with an abnormally thick, viscous, inspissated meconium. The meconium has a reduced water content the result of decreased pancreatic enzyme activity and a prolonged small bowel intestinal transit time. MI can be classified as simple or complicated. Simple MI appears in the first 48 hrs of life with abdominal distension and bilious vomiting. Complicated MI is more severe (< 24 hrs) with progressive abdominal distension, respiratory distress, and peritonitis. X-Ray findings are: dilated bowel loops, absent air-fluid levels, "soap-bubble" granular appearance of distal ileum due to a mixture of air with the tenacious meconium. Therapy consists of Gastrografin enema for simple cases: hyperosmolar solution draws fluid to the bowel lumen causing an osmotic diarrhea. Operative therapy is reserved for failed gastrografin attempts and complicated cases (associated to volvulus, atresias, gangrene, perforation or peritonitis). Surgical procedures has included: ileostomy with irrigation, resection with anastomosis, and resection with ileostomy (Mikulicz and Bishop- Koop). Post-operative management includes: 10% acetylcysteine p.o., oral feedings (pregestimil), pancreatic enzyme replacement, and prophylactic pulmonary therapy. Long-term prognosis depends on the degree of severity and progression of cystic fibrosis pulmonary disease.

Alarming Hemangiomas

Hemangioma is the most common tumor of infancy characterized by proliferation of capillary endothelium. Natural history is of rapid post-natal proliferative growth (8-18 mo) to a slow but inevitable regression during the next 5-8 years (involutive phase). Most are small and harmless. Alarming hemangiomas are associated with heart failure and thrombocytopenic coagulopathy (Kasabach-Merritt syndrome). Management consist of: (1) high dose steroids (rate response 30-60%), (2) radiation therapy, (3) surgery, and recently (4) alpha-interferon.

Alpha-interferon inhibits the proliferation of endothelial cells, smooth muscle cells and fibroblast. Minimal side effects such as fever, elevation of liver

function tests and flu-like syndrome has been reported. Its give subcutaneously, early withdrawal can cause re-growth of the lesion and has been found successful for severe complicating hemangiomas.

CIPO

Chronic Intestinal Pseudo-Obstruction (CIPO) is a rare disorder of intestinal motility in infants and children characterized by recurrent attacks of abdominal pain, distension, vomiting, constipation and weight loss in the absence of obvious mechanical lesions. The disease can be familial or sporadic. Suggested etiology is degeneration of enteric nervous or smooth muscle cells. The diagnosis is based an history, physical exam, radiographies and motility studies. X-Ray hallmarks are: absent strictures, absent, decreased or disorganized intestinal motility, and dilated small/large bowel loops. Associated conditions identified in 10-30% of patients are bladder dysfunction (megacystis) and neurological problems. Histologic pattern portrayed: myenteric plexus hyperplasia, glial cell hyperplasia, and small ganglion cells (hypoganglionosis). Management is primary supportive: intestinal decompression (NG), long-term TPN and antibiotic prophylaxis. Motility agents are unsuccessful. Venting gastrostomy with home parenteral nutrition has shortened the high hospitalization rate associated to this disease process. A similar condition can be seen in early fed prematures due to immaturity of intestinal motility.

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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 Address:http://home.coqui.net/titolugo