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Hirschsprung's Disease:Colostomy ?

Hirschsprung's disease (HD) or the absence of ganglion cells in the distal bowel has traditionally been managed as a three-stage procedure: diagnostic rectal biopsy, leveling colostomy where ganglion cells are present followed by a pull-through (PT) procedure later in life (six months to one year). Historic arguments considered in avoiding a primary neonatal PT were: increased mortality, the limited pelvic size, fragility of neonatal bowel, risk of pelvic nerve damage and injury to muscular sphincters. With the advent of better intensive care support, pathological expertise, adequate instrumentation and technical experience surgeons are managing HD as a single procedure early in life with identical results to the traditional approach. This entails early diagnosis with imaging and suction rectal biopsy, the use of rectal irrigation washout for decompression, and a PT procedure during the same hospitalization. Overall, the open (and recently the laparoscopic) primary PT procedure has shortened the hospital stay, decreased morbidity (that associated with a colostomy) and produce earlier intestinal continuity. This can be accomplished during the first week of life when the weight of the infant is above the four kilograms. Colostomy in the setting of HD will then be needed for cases with: perforation, toxic megacolon, severe enterocolitis, questionable pathology, unavailable frozen section and in the premature infant.

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Typhlitis

Typhlitis (also known as neutropenic enterocolitis) refer to a necrotizing inflammatory process seen in myelosuppressed patients with malignancy (prevalence = 5-9%) who have

chemotherapy-induced intestinal wall damage affecting primarily the ileo-cecal region and ascending colon. Typhlitis is most frequent in patients treated for acute leukemias. Increased intensity of chemotherapeutic regimens may account for a marked increase in the incidence of typhlitis over the past five years. It sometime mimics appendicitis characterized with fever, RLQ abdominal pain, tenderness, nausea, diarrhea and lower GI bleeding. Chemotherapy causes agranulocytosis, intestinal stasis and ischemia with resultant secondary bacteria bowel wall invasion. Typhlitis begin five to 7 days after neutropenia is established. Physical findings are those of abdominal distension and diffuse tenderness. The KUB might show ileus, pneumatosis intestinalis or frank perforation. The CT-Scan demonstrates thickening of the cecal wall, with or without pneumatosis. Most cases can be effectively managed with NG suction, bowel rest, TPN and selective use of antibiotics. Clinical deterioration, failure to improve promptly, persistent peritoneal findings and evidence of pneumatosis are indications for surgery. Right partial colectomy with diverting ileostomy is usually the most appropriate procedure.

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Hepatic Cysts: Sclerotherapy

Benign congenital symptomatic (non-neoplastic) simple hepatic cysts not amenable to surgical therapy can be alternatively managed with ethyl alcohol (ethanol) sclerosis. This can be accomplished with initial ultrasound-guided percutaneous drainage followed by single session ethanol injection of the cyst cavity. The concentration of ethanol should be between 80-95%, at a dose of 10-25% of the cyst volume (never more than 100 cc) applied through the catheter for a short period. During injection and for the next hours monitoring of vital signs, alcohol blood levels, liver function tests and level of consciousness will be needed since some of this alcohol might be absorbed into the blood stream. Minor complications of transient pain, temperature elevation, and hemorrhage into the cyst have been reported. Other series have reported a 75% disappearance rate with minimal morbidity and mortality recommending this as initial therapy for all patients with symptomatic hepatic cysts. Other sclerosants used are tetracycline and doxycycline.

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