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Surgical care of the pediatric Crohn's disease patient

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ABSTRACT

Despite the significant advances in the medical management of inflammatory bowel disease over the last decade, surgery continues to play a major role in the management of pediatric Crohn's disease (CD). While adult and pediatric Crohn's disease may share many clinical characteristics, pediatric Crohn's patients often have a more aggressive phenotype, and the operative care given by the pediatric surgeon to the newly diagnosed Crohn's patient is very different in nature to the surgical needs of adult patients after decades of disease progression. Children also have the unique surgical indication of growth failure to consider in the overall clinical decision making. While surgery is never curative in CD, it has the ability to transform the disease process in children, and appropriately timed operations may have tremendous impact on a child's physical and mental maturation. This monograph aims to address the surgical care of Crohn's disease in general, with a specific emphasis on the surgical treatment of small intestinal and ileocecal involvement.

Introduction

Crohn's disease (CD) is a chronic inflammatory bowel disease caused by a multifactorial etiology of immune, bacterial, and environmental factors in the background of genetically susceptible individuals.[1] The worldwide incidence of CD is rapidly increasing, particularly in children 10–19 years of age, and up to 25% of new CD cases are diagnosed in childhood or adolescence.[2,3] Despite the revolutionary changes that anti-tumor necrosis factor (TNF) agents, such as infliximab, have ushered into the natural history of CD, still one-third of pediatric patients will develop complicated disease such as fistula, stricture, and obstruction and require surgery within five years of their initial diagnosis.[4] There is significant debate as to whether the era of biologics has produced any decrease in the rate of surgical procedures in children.[2,5] Population-based studies have shown that the disease behavior immediately after diagnosis in children tends to be highly inflammatory in nature, with less penetrating and stricturing disease in the early years after initial diagnosis.[4] Because the disease behavior changes over time, pediatric surgeons are likely to perform very different operations on their pediatric CD patients than those required for penetrating disease in patients with CD for decades. Like the adult population, however, surgical care for CD remains palliative, and the insidious threat of short bowel syndrome makes bowel length preservation a surgical priority.

Pediatric surgeons may be called on to treat CD patients during either acute or chronic phases of the disease. Chronic indications fall into four main anatomic categories: ileocecal resections, treatment of small intestinal stricture or fistula, partial or subtotal colectomy for Crohn's colitis, and surgery for perianal complications such as abscess or stricture. This monograph will focus on small intestinal disease, as perianal and colonic Crohn's will be covered separately.

Surgical indications

As medical therapies have drastically improved over the past several decades with the introduction of the anti-TNF agents, the nature of the indications for surgery in CD has changed. Rarely does the patient with small intestinal CD require an emergent operation, although certain absolute indications such as perforation, complete obstruction, or generalized peritonitis may occur. More commonly, the decision for small intestinal resection is made on an elective basis, in concert with the patient, their family, and the pediatric gastroenterologist. It is important to stress that progression to surgical treatment does not imply a failure of medical management, but rather that both medical and surgical expertise is required for the majority of CD patients. Therefore, early surgical consultation should be encouraged, and has the potential to reduce stress, and lessen the uncertainty and anxiety felt by patients and their families regarding what surgical therapy will entail when it becomes necessary.

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Table
Most common acute and chronic surgical indications in small intestinal Crohn's disease.

Acute surgical indications	Chronic surgical indications
Perforation	Stricture
Small bowel obstruction	Enteric fistula
Abscess or phlegmon	Growth retardation
Hemorrhage	Delayed puberty
	Medical treatment complications
	Medical non-compliance

The indications for surgical treatment are as varied as the patient population, but certain surgical principles remain constant. The goal of any operation in pediatric CD is to control the consequences of the stricturing and penetrating inflammatory complications of the disease, while preserving as much bowel length as possible. It should be noted that several known genetic mutations confer a more severe CD phenotype. Notably, mutations in the NOD2/CARD15 gene, which encodes an intracellular receptor to bacterial components on monocytes, confer a phenotype of more severe ileal disease, earlier onset, and higher risk of post-operative recurrence. In the future, medical and surgical treatment may be tailored to patients with genotypes that place them at higher risk for intractable disease. The most common indications for a surgical procedure are shown in the Table.

Surgical emergencies

The need for an acute surgical intervention is relatively uncommon in pediatric CD. The acute presentation of sudden perforation and pneumoperitoneum is very rare, with a more indolent microperforation and abscess/phlegmon formation being a more common clinical scenario.[6] An abscess is most commonly found during an emergency department CT scan obtained for signs and symptoms of fever, leukocytosis, and pain in the setting of CD. Treatment will depend on the location and maturity of the abscess, as well as the ability of the interventional radiologist to provide percutaneous drainage. Drainage, antibiotics, and bowel rest are appropriate in the setting of a stable patient in order to perform a more definitive operation at a later time. Hemodynamically significant hemorrhage is also distinctly uncommon, and in most large series is related to colonic, not small intestinal CD.[6]

In any emergent situation in a Crohn's patient, the abdomen may be hostile with adhesions, fistulas, and inflamed bowel. The goal of emergent laparotomy is to control sepsis, decompress obstructed bowel, and provide the patient with a safe option for resumption of enteral feeds, usually via a diverting ileostomy. Preservation of bowel length should always be a priority, and major resections in the setting of an acute abdomen are not warranted. It should be stressed to patients and their families that ostomies in small intestinal CD are rarely permanent, and stoma formation for 6–8 weeks to control sepsis, treat inflammation, and improve nutrition can allow for safer and less morbid operations than if definitive treatment is attempted in the acute setting.

Special mention must be made regarding obstruction in the setting of capsule endoscopy. Pediatric gastroenterologists are becoming more facile with this diagnostic modality, and it may be used to look for sites of occult bleeding in pediatric CD. By consensus, a capsule is considered retained if it has not passed within 2 weeks of the beginning of the study, and may present with acute small bowel obstruction, or simply a retained foreign body on abdominal radiographs in an otherwise asymptomatic patient.[7] Surgical capsule retrieval may be required when passage is delayed beyond 2 weeks or if the patient presents with pain, vomiting, or other signs of bowel obstruction.

Elective surgical indications

Most surgical procedures performed by pediatric surgeons in the Crohn's population will be elective in nature. The most common indications include strictures, fistulas, and complications of medical therapy. Usually, the decision to proceed with operation can be made after an appropriate diagnostic evaluation, and involves close coordination between surgeon, gastroenterologist, and family. The simple existence of a stricture or fistula is not an indication for operation. Well-documented strictures may be asymptomatic, and surgery should be considered only when significant complications, such as pre-stenotic dilation, weight loss, abdominal distension, or pain develop. Symptomatic fistulas most commonly develop to the urinary bladder, and treatment should involve surgical takedown of the fistula with limited resection of affected bowel, and primary closure of the bladder.[8]

Ileocecal Crohn's disease

Despite the well-known mantra that CD may affect the entire gastrointestinal tract, up to 50% of CD patients will have terminal ileal involvement,[9] and ileocecectomy is one of the most common operations performed for CD in children. A full discussion of the diagnostic workup of a new small intestinal Crohn's disease patient is covered elsewhere in this issue. The typical pediatric patient with ileocecal CD has weight loss, diarrhea, and abdominal pain. Up to one-third of patients will show signs of growth failure as well.[10] A high index of suspicion must be maintained in a patient with this constellation of symptoms. Microcytic anemia, hypoalbuminemia, and elevated erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) are also suggestive. The definitive diagnosis usually depends on endoscopy with biopsies showing granulomatous disease. The upper endoscopy may, however, be normal, and the diagnosis is made by intubation of the terminal ileum and biopsies during colonoscopy. The inability of an experienced endoscopist to reach the terminal ileum, in the proper clinical setting, is also highly suggestive of active ileal CD.

Along with a consistent history, physical exam, and abnormal laboratory studies, cross sectional imaging plays an important role in the diagnosis of terminal ileal stricture. Magnetic resonance enterography (MRE) has become the modality of choice in many centers, given its high accuracy and lack of ionizing radiation.[11] This is especially pertinent given the increasing awareness of the lifetime risk of ionizing radiation, and the high likelihood that CD patients will have multiple imaging studies over their lifetime. MRE can show active inflammation, bowel wall thickening, and pre-stenotic bowel dilatation. It is also useful to evaluate "hot" segments of stricture or disease that are likely to respond to anti-inflammatory therapy, versus "cold" strictures, where the inflammation is quiescent but the bowel remains damaged and obstructive.[12] MRE can also distinguish the patients with the typical ileocecal CD pattern from those with the less common diffuse or more proximal small bowel involvement. A typical example of MRE showing distal ileal Crohn's disease is shown in Figure 1.

Most patients treated in established inflammatory bowel disease centers have had this complete workup before consultation with the pediatric surgeon is sought. The decision to proceed to bowel resection can be a difficult one for the patient and family, and often medical management is continued even in the setting of an identified, fibrostenotic distal ileal stricture. These patients usually have suffered weight loss and post-prandial abdominal