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Surgical management of upper gastrointestinal and small bowel Crohn's disease

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Burill Crohn's convincing description of the disease that now carries his name conceived of the illness as arising exclusively from the terminal ileum, involving other sites only secondarily. As a result, he took the condition to be curable by an adequate operative resection. The current concept is that Crohn's disease may affect any segment of the gastrointestinal tract. The practical implication of this change in thinking is the need to conserve bowel when weighing medical and surgical options for each child. Operations should be used to treat complications of the disease. Absolute indications for the surgery are uncommon and include perforation, bleeding, and refractory obstruction. The margins of resection need only include a short amount of grossly normal intestine. Strictureplasty to relieve obstruction without resection should be done when applicable. Maintenance medication after an operation to limit recurrence or recrudescence is frequently advocated.

In 1932, Burrill Crohn, a gastroenterologist at the Mt. Sinai hospital in New York City, convinced the clinical world of the existence of an inflammatory intestinal disease that now bears his name.[1] His description is striking for the accuracy and breadth of the clinical details. He described patients suffering abdominal pain, diarrhea, fever, anorexia, and anemia. He described complications of the disease, including fistulas, abscesses, and obstruction. He used the term "cobblestone" to depict the gross appearance of the mucosa and recognized the series of transverse ulcerations and deep, longitudinal ulcers that account for the signature appearance of the intervening mucosa. He noted the marked thickening of the intestine from the transmural inflammation. His language is colorful and clear and influenced by the surgeon and the pathologist who contributed to the presentation: "When seen at the operating table, the involved loop is a soggy hoselike mass."

Looking back after 75 years, it appears that Crohn and coauthors surgeon Leon Ginsburg and pathologist Gordon Oppenheimer got very few things wrong. The notable error is that the primary disease arises exclusively from the terminal ileum and that it is curable by an adequate resection. In fact, the authors of the landmark publication planned to call the condition Terminal Ileitis. In the text, the idea is repeated several times that the primary disease arises exclusively from the terminal ileum. The title of the paper, "Regional Ileitis: A Pathologic and Clinical Entity," seems to have been modified at the suggestion of a discussant, Dr. J. A. Bargen from the Mayo Clinic, who commented when the paper was presented: "I am wondering whether the designation 'terminal' is adequately descriptive. To some it has conveyed the meaning of agonal. Perhaps the modifying adjective 'regional' or some other word suggesting its localized nature, instead of the end, would be more suitable." The title was changed, not because the authors doubted that the disease arose exclusively from the terminal ileum, but

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because the term "terminal" could frighten and confuse people who construe it to mean fatal.[2] In the decades since Crohn's contribution, it has been widely recognized that Crohn's disease is an inflammatory condition that can affect every inch of the gastrointestinal (GI) tract from mouth to anus. Crohn and his colleagues thought that adequate resection would cure the disease: "But, in general, the proper approach to a complete cure is by surgical resection of the diseased segment of the small intestine and of the ileocecal valve with its contiguous cecum."[1] Although that remains the most common surgical operation performed for children with Crohn's disease, it clearly and sadly cannot be construed as curative.

I have opened this review by highlighting Crohn's original focus on the terminal ileum for two reasons. First, I wish to address a broader anatomic region, including the foregut and small bowel. More importantly, I would like to emphasize that, in contrast to Crohn's initial concept, the current therapeutic approach to this disease assumes that the entire GI tract is at risk. This modern concept entails several practical implications that I will discuss in this review.

Upper GI Crohn's disease

Histologic evidence by endoscopy of upper GI (UGI) Crohn's disease is quite common in children with Crohn's disease.[3-5] It is rare to encounter Crohn's disease isolated to the foregut without disease located more distally in the GI tract. One study found esophageal, gastric, or duodenal inflammation in 30% of 230 children and adolescents with Crohn's disease.[5] Only 3 of the 230 had Crohn's disease limited to the foregut. The histologic finding of granulomas can be discovered in the foregut of patients with Crohn's disease even in areas that look normal to the endoscopist.[6]

Symptomatic Crohn's disease in this anatomic region is less common. Of 196 children with Crohn's disease studied at the Hospital for Sick Children in Toronto, 10 (5.1%) had symptomatic inflammation in the foregut.[7] All 196 patients had Crohn's elsewhere in the GI tract. Symptoms in the patients with foregut involvement included epigastric pain, weight loss, vomiting, and UGI bleeding in 1 case. None of them had significant obstruction or fistulae. No operations were performed for the disease in the foregut in any of these 10 children. It is difficult to know from this study whether the symptoms were actually due to proximal inflammation since many children also had distal involvement. It is also unclear whether the foregut inflammation was necessarily due to Crohn's disease since only 3 of the 10 patients had documented granulomas.

In the unusual case in which operative intervention is required for Crohn's disease of the foregut, obstruction is by far the most common indication. Fistulas rarely occur in gastroduodenal Crohn's disease. Hemorrhage is usually chronic and difficult to distinguish from peptic ulcer disease. The preferred surgical treatment for obstructive gastroduodenal Crohn's disease is the gastrojejunostomy bypass procedure. It is uncertain whether vagotomy should be done in conjunction with gastrojejunostomy to prevent marginal ulceration. The number of reported cases is too small for a definitive recommendation, and the question has not been carefully studied. Delayed gastric emptying is a potential risk in this situation, and vagotomy may exacerbate this problem. Resection for a Crohn's stricture in this anatomic area has few advocates because of the associated morbidity.

Duodenoplasty to widen the strictured duodenum has been attempted with limited success. In 1 report of 13 such cases, there was a high rate of recurrence, persistence, and surgical complications: the obstruction persisted in 1/13, recurred in 6/13, and the suture line broke down in 2/13.[8] The Cleveland Clinic reported 34 patients who underwent surgery for duodenal Crohn's disease with better results.[9] All but one was obstructed. Gastrojejunal bypass was done in 21 patients and strictureplasty in 13 patients. One patient in each group required re-operation. Two surgical complications occurred in each group. Vagotomies were done in 16 of the 21 patients undergoing bypass and, interestingly, in 5 of the 13 undergoing strictureplasty. Gastrostomy was added in 3 of the 21 who underwent bypass and 6 of the 13 who underwent strictureplasty.

Rare fistulas involving the stomach or duodenum usually result from primary disease elsewhere, affecting the foregut as the innocent bystander. As with fistulas between loops of small bowel, or from small bowel to colon, bladder, or vagina, the surgeon should resect the site from which the fistula arises and oversew the innocent end.

Evidence of esophageal inflammation (and occasionally granulomas) of the esophagus is not uncommon in children with Crohn's disease. However, symptomatic esophageal disease is rare and fortunately never requires operative treatment.

Small bowel Crohn's disease

It is ironic that the disease originally thought to arise exclusively from the terminal ileum can be said to affect the colon more than any other site in the GI tract when cases of terminal ileitis with colonic involvement are included. When *Seminars in Pediatric Surgery* last reviewed this subject, Telander and Schmeling documented the location of the disease in 177 children followed by the Mayo Clinic. Sites of involvement were often multiple, and when all sites of colonic involvement are combined, they outnumber those sites with disease in the small bowel alone. Nonetheless, the terminal ileum was involved in 76.8% of the patients, many of whom also had involvement of the colon[10] (Figure 1).

With clear recognition that Crohn's disease is not cured by resection, surgery is reserved for patients who fail medical management or develop debilitating complications. The most common indications for surgery in pediatric Crohn's