Practice Parameters

Practice Parameters for the Surgical Management of Crohn's Disease

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T he American Society of Colon and Rectal Surgeons is dedicated to ensuring high-quality patient care by advancing the science, prevention, and management of disorders and diseases of the colon, rectum, and anus. The Standards Committee is composed of Society members who are chosen because they have demonstrated expertise in the specialty of colon and rectal surgery. This Committee was created to lead international efforts in defining quality care for conditions related to the colon, rectum, and anus. This is accompanied by developing Clinical Practice Guidelnes based on the best available evidence. These guidelines are inclusive and not prescriptive. Their purpose is to provide information on which decisions can be made, rather than dictate a specific form of treatment. These guidelines are intended for the use of all practitioners, health care workers, and patients who desire information about the management of the conditions addressed by the topics covered in these guidelines.

It should be recognized that these guidelines should not be deemed inclusive of all proper methods of care or exclusive of methods of care reasonably directed to obtaining the same results.

The ultimate judgment regarding the propriety of any specific procedure must be made by the physician in light of all of the circumstances presented by the individual patient.

DISEASE CLASSIFICATION

A World Congress of Gastroenterology Working Party developed the Vienna Classification to help standardize the categorization of Crohn's disease.[1] The group prospectively designed this simple phenotypic classification system, which is based on objective and reproducible clinical variables, which include age at diagnosis, anatomic location, and disease behavior. The age at diagnosis is grouped into patients younger than aged 40 years and those aged 40 years and older. The anatomic location is stratified as terminal ileum, colon, ileocolon, and upper gastrointestinal tract. Terminal ileal disease is disease limited to the lower third of the small bowel with or without cecal involvement. Colon disease is any colonic involvement between the cecum and rectum without small-bowel disease. Ileocolon disease is disease of the terminal ileum with colonic involvement. Upper gastrointestinal disease is any disease located proximal to the terminal ileum regardless of involvement in other areas. The disease behavior is classified as nonstricturing, nonpenetrating (inflammatory), stricturing, and penetrating. Management principles for perianal Crohn's disease have been discussed in a separate practice parameter.[2]

OPERATIVE INDICATIONS

Failed Medical Therapy

1. Patients with disease-related symptoms who suffer from medically unresponsive disease, demonstrate an inadequate response, manifest medication-related complications, or appear noncompliant with medication should be considered for operation. Level of Evidence: II; Grade of Recommendation: B.

The therapeutic options for symptomatic intestinal disease are primarily predicated upon disease severity and response to previous therapies. Patients with mild-to-moderate disease are able to tolerate oral alimentation without symptoms or signs of abdominal tenderness, dehydration, obstruction, painful mass, toxicity, or >10 percent weight loss.[3] This lesser level of disease is typically managed with first-line therapy, including aminosalicylates, antibiotics, and steroids. If these medications are unsuccessful, the disease is considered severe.[4] Overall, steroids are ineffective in 20 to 30 percent of patients, and 20 to 45 percent of initial responders will relapse with steroid tapering or withdrawal.[5–7] Accordingly, both purine analogs and methotrexate can be used because they may successfully treat steroid-refractory or steroid-dependent disease.[4,8] Alternatively, infliximab and natalizumab are useful in many patients with severe disease that is refractory to steroids, whereas cyclosporine, tacrolimus, mycophenolate mofetil, and elemental diets have been anecdotally beneficial.[4,8] Surgical treatment is warranted if these first-line and second-line therapies fail to induce remission safely in severe disease states, but surgery also should be considered before escalating medical therapy for patients with severe or steroid-dependent disease that it limited in disease extent, particularly in individuals with stricturing behavior or those with contraindications or risk factors for further medical therapy.[9]

Perforation

1. Patients with symptoms and/or signs of free perforation should undergo operation. Level of Evidence: III; Grade of Recommendation: B.

Patients with Crohn's disease can present with free perforation of the small or large bowel; perforation is the indication for surgery in 1 to 16 percent of instances.[10–15] Immediate resection of the perforated segment is preferred over simple suture closure because of a relatively high mortality rate associated with the latter.[11] After resection of a small-bowel perforation, an end stoma, diverted anastomosis, or nondiverted anastomosis can be performed depending on the presentation and operative findings.[13–18] Colorectal perforations are more commonly resected in combination with a proximal stoma and mucous fistula or Hartmann's closure of the distal bowel;[10,11,13,15] alternatively, a diverted anastomosis can be considered.

2. Patients with large enteroparietal, interloop, intramesenteric, or retroperitoneal abscesses may be managed by antibiotics and percutaneous drainage. If this approach is unavailable or unsuccessful, the

LEVELS OF EVIDENCE AND GRADE RECOMMENDATION

Level	Source of Evidence
I	Meta-analysis of multiple well-designed, controlled studies, randomized trials with low-false positive and low-false negative errors (high power)
II	At least one well-designed experimental study; randomized trials with high false-positive or high false-negative errors or both (low power)
III	Well-designed, quasi-experimental studies, such as nonrandomized, controlled, single-group, preoperative–postoperative comparison, cohort, time, or matched case-control series
IV	Well-designed, nonexperimental studies, such as comparative and correlational descriptive and case studies
V	Case reports and clinical examples
Grade	Grade of Recommendation
A	Evidence of Type I or consistent findings from multiple studies of Type II, III, or IV
B	Evidence of Type II, III, or IV and generally consistent findings
C	Evidence of Type II, III, or IV but inconsistent findings
D	Little or no systematic empirical evidence

Adapted from Cook DJ, Guyatt GH, Laupacis A, Sackett DL. Rules of evidence and clinical recommendations on the use of antithrombotic agents. Chest 1992;102(4 Suppl):305S–311S. Sackett DL. Rules of evidence and clinical recommendations on the use of antithrombotic agents. Chest 1989;92(2 Suppl):2S–4S.

patient should undergo surgical drainage with or without resection. Level of Evidence: III; Grade of Recommendation: B.

Intra-abdominal abscesses in patients with Crohn's disease typically result from a perforation that is contained by surrounding structures, and this may include segments of nondiseased bowel. Antibiotics and percutaneous drainage of a large (≥5 cm) abscess usually controls the sepsis,[19–22] uncommonly results in an enterocutaneous fistula,[23] and occasionally obviates the need for future surgery.[20,22–25] Conversely, operative drainage is much more likely to cause an enterocutaneous fistula that requires later surgery.[26] Surgical eradication of the abscess generally requires excision of the diseased bowel responsible for the abscess and possibly the nondiseased bowel that has quarantined the sepsis. In this setting, removal of the diseased and nondiseased bowel risks immediate or future short bowel syndrome.[23]

3. Patients with enteric fistulas and symptoms or signs of localized or systemic sepsis that persist despite appropriate medical therapy should be considered for operation. Asymptomatic patients with internal fistulas do not typically require surgery. Level of Evidence: III; Grade of Recommendation: B.

Fistulas originating in diseased bowel and secondarily involving other intra-abdominal organs or the skin are not usually associated with localized or systemic sepsis. If sepsis is present, the patient should be started on broad-spectrum antibiotics and imaging studies should be performed to exclude a concomitant abscess,[27] which should be drained as previously described. Regardless of whether an abscess is present, persistent sepsis usually warrants excision of the diseased bowel.[28] Management of the target or "innocent bystander" organ is based on whether it is diseased bowel, noninflamed bowel, or another internal organ. Diseased bowel is generally resected, noninflamed bowel can be primarily closed, and other internal organs (e.g., bladder, vagina) can be primarily closed or left to heal by secondary intention.[29–34] Surgery is usually not necessary if sepsis and symptoms are absent.[35–38]

Obstruction

1. Patients with symptomatic strictures in any location that do not appear amenable or responsive to medical therapy should undergo operation. Level of Evidence: III; Grade of Recommendation: B.

The incidence of fibrostenosing lesions in patients with Crohn's disease increases with longer disease duration, and this behavior is most commonly seen in patients with upper gastrointestinal disease involvement.[39,40] Computed tomography and magnetic resonance enterography can sometimes help discern whether these areas of narrowing have an inflammatory component that might respond to medical therapy.[41–44] If the patient is symptomatic and the stricture appears fibrotic with minimal inflammation, or if medical therapy fails to adequately improve the symptoms, surgery is generally warranted.

2. Patients with asymptomatic strictures of the colon that cannot be adequately surveyed by biopsy and/or cytology brushing should be considered for operation. Level of Evidence: III; Grade of Recommendation: B.

Approximately 7 percent of large bowel strictures in patients with Crohn's disease are malignant,[45] and consequently they should be surveyed for neoplastic transformation. This is usually performed with multiple biopsies and cytologic brushing. If the stricture cannot be appropriately surveyed or if neoplastic changes are identified, resection is usually warranted.

Inflammation

1. Patients with acute colitis and symptoms or signs of impending or actual perforation should undergo operation. Level of Evidence: III; Grade of Recommendation: B.

Patients with Crohn's disease and severe or fulminant colitis are at risk for toxic megacolon and perforation similar to persons with ulcerative colitis. Severe colitis is generally defined by the Truelove and Witts[46] criteria, which include >6 bloody bowel motions per day, fever (temperature >37.5°C), tachycardia (heart rate >90 beats per minute), anemia (hemoglobin <75 percent of normal), and elevated sedimentation rate (ESR >30 mm per hour). Alternatively, toxic or fulminant colitis is characterized by more than ten bloody bowel motions per day, fever (temperature >37.5°C), tachycardia (heart rate >90 beats per minute), anemia (transfusion required), elevated sedimentation rate (ESR >30 mm per hour), colonic dilatation on radiography, and abdominal distention with tenderness.[47]

Although supporting evidence is limited, severe or fulminant colitis should be treated in the same way,

regardless of the underlying inflammatory bowel disease.[48–51] An abdominal film that reveals transverse colon distention >6 cm[52] or persistent gaseous distension in a colonic segment[53] indicates toxic megacolon and suggests that the patient is at risk for perforation. Persistent or progressing colonic dilatation, pneumatosis coli, evolving local peritonitis, and multiple organ failure also are evidence of impending or actual perforation[54–56] and surgery is warranted.

2. Patients with acute colitis whose condition worsens despite appropriate medical therapy or fails to significantly improve after 48 to 96 hours of medical therapy should be considered for operation. Level of Evidence: III; Grade of Recommendation: B.

Again the experience with ulcerative colitis is extrapolated to Crohn's disease, and patients are judged to have failed medical therapy if their condition worsens while on medical therapy or if their condition fails to improve after a period of initial stabilization. The need for and timing of surgery in patients whose condition initially improves, but then plateaus, is sometimes difficult to determine. Continued observation of these patients may exhaust their physiologic reserve without benefit. Thus, some authors recommend an observation period of 48 to 96 hours, after which surgery is indicated if the patient fails to improve.[54,57] Evidence specifying the most appropriate period for medical therapy, however, is lacking.

Hemorrhage

1. Patients with massive hemorrhage originating from any location may be managed by interventional radiologic and/or endoscopic techniques. If the patient is too unstable for this, or if this approach is unavailable or unsuccessful, the patient should undergo operation. Level of Evidence: III; Grade of Recommendation: B.

Massive gastrointestinal hemorrhage is an uncommon event in patients with Crohn's disease. Other common etiologies of gastrointestinal hemorrhage, such as peptic ulcer disease and gastritis, should be excluded. If the hemorrhage is a direct consequence of Crohn's disease, it most commonly occurs from an ulcer that has eroded into a submucosal vessel. If the patient is stable and the source of bleeding can be endoscopically identified, local measures can be used to halt the bleeding.[58–61] If the bleeding is too brisk to permit adequate endoscopic visualization or the source of hemorrhage cannot be identified, mesenteric angiography can be attempted and the

bleeding site might be embolized.[58,61,62] Patients who are hemodynamically unstable or fail to respond to these measures usually require laparotomy, sometimes with intraoperative endoscopy, and resection of the responsible bowel segment.[63,64]

Neoplasia

1. Patients with long-standing Crohn's disease of the ileocolon or colon should undergo endoscopic surveillance. Level of Evidence: III; Grade of Recommendation: B.

Patients with Crohn's disease are at increased risk for malignancy at the sites of intestinal inflammation. Although few studies have adequately investigated this issue, the risk for large bowel cancer seems to be similar to that seen with ulcerative colitis.[65,66] Consequently, patients with colitis often are advised to undergo a screening colonoscopy after eight to ten years of disease symptoms, and surveillance endoscopy every one to two years thereafter.[67] Ideally, surveillance colonoscopy should be performed when the disease is in remission to minimize confusion interpreting neoplastic changes. Four quadrant random biopsies at 10-cm intervals along the colon and rectum may be obtained, as well as biopsies of any strictures, lesions, or mass; pseudopolyps that do not appear suspicious need not be sampled. Adenomatous-appearing polyps should be removed if possible, and the adjacent flat mucosa should be biopsied to exclude associated dysplasia.

2. Patients with carcinoma, dysplasia-associated lesion or mass (DALM), high-grade dysplasia, or multifocal, low-grade dysplasia of the colon or rectum should undergo resection. Level of Evidence: III; Grade of Recommendation: B.

Patients with Crohn's disease enrolled in a surveillance program have a 22 percent probability of developing dysplasia or cancer by the fourth surveillance examination, after a negative initial screening colonoscopy.[68]

Dysplasia may be complicated by concomitant or future invasive cancer, and therefore resection is appropriate if a dysplasia-associated lesion or mass, high-grade dysplasia, or multifocal, low-grade dysplasia of the colon or rectum is identified and confirmed.[65,69] The appropriate extent of the resection is unclear and could range from a limited segment that includes only the inflamed bowel to the entire colon and rectum. The optimal management of low-grade dysplasia in flat mucosa also is uncertain. In a small

observational study, lack of progression was noted in all six patients with flat low-grade dysplasia during a 17.8-year period of follow-up that included an average of three surveillance colonoscopies.[70] Controversy also surrounds the natural history and management of adenoma-like DALMs. Specifically, one report suggests that adenoma-like DALMs can be removed effectively by colonoscopic resection without risk of future dysplasia or carcinoma,[71] whereas another reported a 22 percent incidence of progression.[70]

3. Patients with long-standing Crohn's disease of the terminal ileum, ileocolon, or upper gastrointestinal locations should undergo biopsy of suspicious lesions at the time of strictureplasty. Level of Evidence: III; Grade of Recommendation: B.

Small-bowel adenocarcinoma has been reported at the site of previous strictureplasty,[72–75] and consequently the stricture site should be carefully inspected and any suspicious areas should be biopsied.

Growth Retardation and Extraintestinal Manifestations

1. Prepubertal patients with significant growth retardation despite appropriate medical therapy should be considered for operation. Level of Evidence: IV; Grade of Recommendation: C.

Approximately one-quarter of patients with Crohn's disease have disease onset before age 18 years, and growth failure often is present at the time of diagnosis.[76] The typical pattern is growth retardation associated with impaired skeletal maturation and delayed puberty. Many pediatric patients demonstrate a subnormal height velocity, and half of these will display short stature.[77] Although the medical treatment of Crohn's disease in the pediatric population is shifting toward a more aggressive approach, adequate growth often fails to occur despite medical therapy or supplemental enteral nutrition.[78,79] In these instances, surgery can restore normal growth, but it is optimally performed before the onset of puberty.[80–84]

2. Patients with symptomatic disorders of the skin, mouth, eye, or joints who fail to respond to medical therapy should be considered for operation. Level of Evidence: IV; Grade of Recommendation: C.

Extraintestinal manifestations (EIMs) of Crohn's disease occur in up to 30 percent of patients.[85–87] The presence of one EIM sometimes predisposes an individual to the development of others. Some EIMs are temporally related to intestinal disease activity, whereas others occur independently. Erythema nodosum, oral aphthous ulcers, episcleritis, and some types of peripheral arthritis tend to parallel the intestinal disease activity, whereas pyoderma gangrenosum, uveitis, spondyloarthropathy, and primary sclerosing cholangitis do not. For the former group, therapy for the EIM focuses on treatment of the underlying intestinal disease, and medical management is usually sufficient. Nevertheless, if medical therapy fails to adequately control these manifestations, resection of the diseased intestine is warranted. For the group of EIMs that behave independent of the intestinal disease, surgery has no role in the management of the EIM.

SITE-SPECIFIC OPERATIONS

Terminal Ileum, Ileocolon, and Upper Gastrointestinal Tract

1. Patients who require surgery for disease of the jejunum, proximal ileum, terminal ileum, or ileocolon without existing or impending short-bowel syndrome should usually undergo resection of the affected bowel. Level of Evidence: III; Grade of Recommendation: B.

Resection continues to be the most commonly performed operation for symptomatic penetrating or inflammatory disease affecting the small bowel and proximal colon, especially when existing or impending short-bowel syndrome is unlikely. The procedure can be performed through a laparoscopic or an open approach because the two methods are associated with comparable short-term outcomes, such as operative blood loss and rates of wound infection, anastomotic leak, and intra-abdominal abscess when performed for uncomplicated ileocolic disease. The laparoscopic mode is associated with a longer operative time but a shorter length of stay and more rapid resolution of ileus.[88] Regardless of the approach, limited disease-free (2 cm) resection margins are adequate, because they conserve bowel length and are not associated with an increased risk of disease recurrence.[89] The anastomosis can be constructed with suture or mechanical stapling devices. Although controversial, some reports conclude that a stapled anastomosis is safer assuming the bowel wall is not thickened,[90–92] and a larger side-to-side anastomosis may provide a lower risk of recurrence.[90,93–96]

Most patients with penetrating or inflammatory disease of the terminal ileum or ileocolon can

undergo resection of the offending bowel without significant risk to nondiseased bowel, superior mesenteric vessels, or retroperitoneal structures. Nevertheless, in rare cases when there is concern about damage to these structures, the diseased segment may be bypassed by using an ileocolostomy or proximal loop ileostomy. At the same time, any septic focus should be drained. Definitive resection at a later date is recommended, because the bypassed segment is at risk for recurrent disease[97] and later adenocarcinoma.[98] The subsequent operation should be delayed for several months to allow resolution of the inflammatory process and a safer excision.

Certain factors are associated with an increased risk for anastomotic dehiscence. Although studies do not universally agree, these factors include: long-term preoperative steroid use,[99] impaired preoperative nutritional status,[100] low serum albumin (<3 mg/dl),[101] low hemoglobin,[102] urgent surgery,[100] and abscess or fistula at the time of laparotomy.[101] Preoperative immunomodulators (*i.e.,* purine analogues, methotrexate) or biologic agents (*i.e.,* infliximab) do not seem to increase the risk of septic complications.[103,104]

2. Patients who require surgery for nonphlegmonous strictures of the jejunum, ileum, or ileocolon, and existing or impending short bowel syndrome should typically undergo strictureplasty. Level of Evidence: III; Grade of Recommendation: B.

Strictureplasty should be strongly considered in patients with multiple strictures of the jejunum, proximal ileum, or terminal ileum. This procedure can be performed by using a variety of techniques, depending on the length of the affected bowel[105–111] and often is performed in conjunction with resection. The morbidity[112,113] and recurrence rate[112–114] are equivalent to resection. Similarly, patients with nonphlegmonous strictures of the ileocolon and existing or impending short-bowel syndrome should be considered for strictureplasty because the operation is safe and effective.[111,112,115–117]

3. Patients with symptomatic disease of the stomach or duodenum should be considered for bypass of the affected area or strictureplasty. Level of Evidence: III; Grade of Recommendation: C.

Bypass options for refractory obstruction or pain secondary to gastroduodenal Crohn's disease include gastrojejunostomy and duodenojejunostomy. Truncal vagotomy with gastrojejunostomy may decrease the risk of marginal ulceration but increases the likelihood of diarrhea. Highly selective vagotomy may

avoid these problems. Compared with a bypass procedure, resection is associated with prohibitive morbidity.[118] Whereas symptomatic strictures of the distal stomach or duodenum can be treated with bypass, nonperforated, nonphlegmonous stenotic lesions in this region also can be safely managed by strictureplasty.[119,120] The operative morbidity and recurrence rates are comparable to bypass,[121,122] and strictureplasty avoids the need to consider vagotomy.

4. Patients with symptomatic, accessible strictures of the intestinal tract can be considered for endoscopic dilatation of the affected area. Level of Evidence: II; Grade of Recommendation: C.

Mild-to-moderate stricturing disease of the terminal ileum, colon, or duodenum can be safely and effectively treated by endoscopic dilatation with[123] or without[124–135] concomitant steroid injection. The procedure allows surgery to be postponed or avoided in many patients with Crohn's disease and short intestinal strictures. However, recurrent symptoms frequently necessitate a repeat procedure, and a dilatation should not be performed unless surgical services are available to intervene if perforation occurs.

Colon

1. Patients with disease of the colon that requires emergency or urgent surgery should typically undergo subtotal or total colectomy with end ileostomy. Level of Evidence: III; Grade of Recommendation: B.

Subtotal or total colectomy with end ileostomy and Hartmann's closure of the distal bowel or creation of a mucous fistula is a safe procedure in patients with severe or fulminant colitis who require surgery.[136,137]

This approach removes most of the inflamed intestine with a relatively simple operation that avoids a pelvic dissection and an anastomosis and can be performed through a laparoscopic or open approach.[138–140] Extrafascial placement of the closed rectosigmoid stump may be associated with fewer pelvic septic complications than an intraperitoneal position.[141,142] Transanal drainage of the distal stump may further decrease the risk of pelvic sepsis.[143]

2. Patients with disease of the colon that requires elective surgery may undergo segmental or total colectomy with or without a primary anastomosis. Level of Evidence III; Grade of Recommendation: B

Symptomatic disease of the colon with rectal sparing can be managed by removal of only the diseased segment or by total colectomy. The oper-

ations are comparable with respect to operative complications, need for a permanent stoma, and risk of recurrence, but segmental resection is associated with earlier recurrence.[144] Total colectomy with ileoproctostomy is preferable if two or more colonic segments are affected, because this subgroup has a higher recurrence rate when segmental resections are performed.[144]

3. Patients who require surgery for disease of the rectum may undergo total proctocolectomy or proctectomy with creation of a stoma. Level of Evidence: III; Grade of Recommendation: C.

Refractory proctitis usually warrants total proctocolectomy with a permanent ileostomy,[145–147] but proctectomy alone with creation of a colostomy can be performed if the colon is spared.[148] Usually the entire rectum should be excised because cancer has been found in patients with even a short Hartmann's remnant.[149] A few centers have suggested that a restorative proctocolectomy with ileal pouch-anal anastomosis can be considered in this situation if the small bowel is unaffected and there is no perianal disease.[150,151] However, many have reported a poor outcome in patients with Crohn's disease who underwent this procedure.[152–155]

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