Clinical Practice Guideline for the Surgical Management of Crohn’s Disease

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Prepared on behalf of The Clinical Practice Guidelines Committee of the American Society of Colon and Rectal Surgeons

The 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 Clinical Practice Guidelines 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 accomplished by developing Clinical Practice Guidelines based on the best available evidence. These guidelines are inclusive, and not prescriptive. Their purpose is to provide information based 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 the circumstances presented by the individual patient.

STATEMENT OF THE PROBLEM

Crohn’s disease is a chronic, unremitting, incurable inflammatory disorder that can affect any segment of the intestinal tract, as well as extraintestinal sites. Disease behavior is classified as nonstricturing, nonpénetrating (ie, inflammatory), fibrostenotic, or penetrating, and is prone to change over the course of the disease. Approximately 19% to 38% of patients have stricturing or penetrating complications at the time of diagnosis, whereas 61% to 88% of patients will have developed these manifestations after 20 years of disease. Although the etiology of Crohn’s disease remains unknown, the disease likely results from a complex interplay between conditioning factors (eg, genetics, triggering events) and effector mechanisms that lead to a dysregulation of both intestinal immune and nonimmune functions.

Before the introduction of anti-tumor necrosis factor (TNF) medications, population-based cohorts from Canada, Denmark, Norway, Sweden, the United States, and Wales reported that surgical intervention is performed in 27% to 61% of patients with Crohn’s disease within 5 years of diagnosis. Independent risk factors associated with undergoing an initial surgery (versus reoperation) according to similar population-based cohorts include current tobacco usage, disease location, stricturing or penetrating disease behavior, and (although somewhat controversial) the early use of high-dose glucocorticoids or immunomodulators. This clinical practice guideline will focus on the surgical management of patients with Crohn’s disease.

METHODOLOGY

This clinical practice guideline is based on the previous American Society of Colon and Rectal Surgeons’ Practice Parameters for the Surgical Management of Crohn’s disease that was published in 2007. The management of perianal disease in patients with Crohn’s disease is discussed in a separate clinical practice guideline. An organized search of MEDLINE, PubMed, EMBASE, and the Cochrane Database of Collected Reviews was performed from 1978 through February 2015. Key-word combinations included “Crohn’s disease”; “colitis”; “ileitis”; “fibrostenotic”; “structure”; “abscess”; “phlegmon”; “fistula”; “megacolon”; “fulminant”;

Directed searches of the embedded references from the primary articles were also performed in selected circumstances. Although not intended to be exclusionary, the authors primarily focused on English language articles and studies in adults. Recommendations were formulated by the primary authors and reviewed by the entire ASCRS Clinical Practice Guidelines Committee. The final grade of recommendation was performed by using the GRADE system (Table 1).13

OPERATIVE INDICATIONS

Failed Medical Therapy

1. Patients who demonstrate an inadequate response to, develop complications from, or are noncompliant with medical therapy should be considered for surgery.

Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

The goals of medical treatment of Crohn’s disease are to achieve remission in patients with active disease and to avoid relapse in those with quiescent disease. Clinical evidence of improvement usually begins in approximately 2 to 4 weeks for the majority of classes of medications used in patients with Crohn’s disease. For immunomodulators, a plateau in improvement of clinical symptoms is demonstrated within 12 to 16 weeks.14 Patients achieving clinical remission should then be considered for maintenance medical therapy.

Surgical intervention is often warranted in patients who are unable to tolerate medical therapy because of side effects,15,16 are noncompliant with medical therapy, or do not adequately control their disease activity with medical therapy.17 One special circumstance involves those patients requiring steroids, where regardless of disease severity, steroids should be transitioned to another medication as soon as possible owing to the deleterious effects of chronic glucocorticoid use. If a patient cannot tolerate medical therapy other than steroids,18 or if their disease is limited in extent (eg, short-length ileocolic disease), surgery should be strongly considered.

2. Patients receiving therapy with anti-TNFs, high-dose glucocorticoids, and/or cyclosporine may warrant staged procedures because of concerns about postoperative complications; however, decisions should be individualized based on the patient’s risk stratification, overall clinical status, and surgeon judgment. Grade of Recommendation: Weak recommendation based on low- or very low-quality evidence, 2C.

The association between antitumor necrosis factor agents and complications in patients with Crohn’s disease remains unclear, with studies reporting disparate results.15–30 Unfortunately, methodological flaws in the individual studies, ranging from a failure to control for disease activity, being underpowered, or having significant heterogeneity, make it difficult to provide widespread recommendations. The pharmacokinetics of monoclonal antibodies such as infliximab allow for persistent therapeutic concentrations for at least 8 weeks after infusion. Given this controversy, elective surgeries in patients with minimal symptoms may be accordingly delayed as permitted until the end of this period or longer when feasible. However, in both the elective and emergent setting, the decision to perform a staged procedure and/or diversion should involve the patient’s overall clinical picture and risk stratification. Factors include nutritional status, intraoperative factors (ie, site of resection, technical issues, associated inflammation), hemodynamic status, and the presence or absence of a surgical delay following failure to respond to medical therapy.39–42

Similarly, the impact of preoperative high-dose glucocorticoid therapy (eg, prednisone >20 mg per day for >6 weeks) on postoperative complications is controversial.43–47 Steroids appear to increase the risk for infectious complications, especially when given in combination with anti-TNF agents or cyclosporine.18,46,47 This potential risk should be balanced against that of diversion and its inherent complications.50 The need for surgery and the timing of operative intervention in patients whose condition initially improves, but never completely normalizes, should be left to the clinical judgment of the treating physician. Continued observation of these patients may exhaust their physiological reserve without benefit, and operative intervention is often recommended despite initial improvement – such as in the case of using cyclosporine to salvage patients with severe colitis. Unfortunately, the literature regarding cyclosporine is principally limited to patients with ulcerative colitis, although this group of patients may be analogous to patients with Crohn’s colitis.51–63

Inflammation

1. Patients with acute colitis who have symptoms or signs of impending or actual perforation should typically undergo surgery. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Acute severe colitis is generally defined as ≥6 grossly bloody bowel movements per day plus 1 sign of systemic toxicity,
TABLE 1. The GRADE system-grading recommendations

<table>
<thead>
<tr>
<th>Description</th>
<th>Benefit vs Risk and Burdens</th>
<th>Methodological Quality of Supporting Evidence</th>
<th>Implications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>Strong recommendation, High-quality evidence</td>
<td>Benefits clearly outweigh risk and burdens or vice versa</td>
<td>RCTs without important limitations or overwhelming evidence from observational studies</td>
</tr>
<tr>
<td>1B</td>
<td>Strong recommendation, Moderate-quality evidence</td>
<td>Benefits clearly outweigh risk and burdens or vice versa</td>
<td>RCTs with important limitations (inconsistent results, methodological flaws, indirect or imprecise) or exceptionally strong evidence from observational studies</td>
</tr>
<tr>
<td>1C</td>
<td>Strong recommendation, Low- or very low-quality evidence</td>
<td>Benefits clearly outweigh risk and burdens or vice versa</td>
<td>Observational studies or case series</td>
</tr>
<tr>
<td>2A</td>
<td>Weak recommendation, High-quality evidence</td>
<td>Benefits closely balanced with risks and burdens</td>
<td>RCTs without important limitations or overwhelming evidence from observational studies</td>
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<td>2B</td>
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</tr>
<tr>
<td>2C</td>
<td>Weak recommendation, Low- or very low-quality evidence</td>
<td>Uncertainty in the estimates of benefits, risks, and burden; benefits, risk, and burden may be closely balanced</td>
<td>Observational studies or case series</td>
</tr>
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GRADE = Grades of Recommendation, Assessment, Development, and Evaluation; RCT = randomized controlled trial.

which include anemia (<10.5 g/dL), elevated erythrocyte sedimentation rate (>30 mm/h), fever (>37.8°C), and tachycardia (>90 beats per minute). Alternatively, “fulminant” colitis is characterized by >10 bloody bowel movements per day, daily continuous bleeding, a blood transfusion requirement, an elevated erythrocyte sedimentation rate (>30 mm/h), fever (>37.5°C), tachycardia (>90 beats per minute), abdominal tenderness and distension, and colonic dilation on abdominal radiographs.

Toxic megacolon is defined as a total or segmental nonobstructive dilation (ie, transverse colon >5.5 cm in the midline) of the colon associated with systemic toxicity, and it suggests that the patient is at higher risk for perforation. When severe colitis exists in the setting of persistent or progressing colonic dilation, evolving local peritonitis, and multiple organ failure, impending or actual perforation is of even greater concern and surgery is typically warranted. Outcomes associated with the medical management of severe or fulminant colitis secondary to Crohn’s disease are relatively lacking, but the experience gained from treating patients with severe ulcerative colitis is again generally extrapolated. Alternative causes of severe colitis (eg, Clostridioides difficile and cytomegalovirus) should be excluded with stool studies and endoscopic biopsy. Endoscopy with minimal insufflation may be used with extreme caution to assess for concomitant disease as well as the endoscopic severity of disease, which is associated with the likelihood for response to medical therapy. Medical therapy with parenteral glucocorticoids or biological agents may be initiated, whereas narcotics and antidiarrheal agents should generally be avoided to prevent progression to megacolon. Bowel rest and parenteral antibiotics have not proven to reliably improve severe or fulminant colitis, although their use should be dictated by the individual patient’s clinical scenario. Medical therapy is judged to have failed in patients when their symptoms are uncontrolled by medications or there is clinical evidence of persistent or progressive disease (eg, persistent tachycardia, hypotension, fever, leukocytosis, or persistent elevation of inflammatory markers such as C-reactive protein). Emerging evidence indicates that therapeutic drug level and anti-TNF antibody assessments are also useful in the determination of medical therapy success or failure. Patients receiving parenteral glucocorticoids should typically show improvement within 2 to 3 days after initiating therapy. If not, salvage therapy with a biological agent or operative intervention is often required. As seen in ulcerative colitis, a favorable response to a biological agent should normally be demonstrated within 5 to 7 days after receiving the medication. Failure to respond, again, typically indicates a need for operative intervention.
Stricture

1. Endoscopic dilation may be considered for patients with symptomatic small-bowel or anastomotic strictures that are not amenable to medical therapy. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Strictures complicating Crohn’s disease can arise anywhere in the intestinal tract, and they are conventionally classified as either inflammatory or fibrostenotic in nature. However, these categories are not mutually exclusive: intestinal fibrosis is a dynamic process, and inflammation and fibrosis can readily coexist within the same stricture or region.\textsuperscript{69} Computed tomography enterography and magnetic resonance enterography can diagnose small-bowel strictures with a high level of accuracy, and may be considered apart or in addition to standard CT.\textsuperscript{70,71} Both CT enterography and MR enterography have a relatively high sensitivity and specificity for identifying disease activity that helps discriminate between inflammatory and fibroptic strictures.\textsuperscript{72} However, MRI is not associated with radiation exposure like CT imaging, and may be more appropriate in younger patients who potentially face an increased lifetime cumulative radiation exposure. At present, neither modality is recommended for the routine assessment of ileocolic or colonic strictures unless the stricture or more proximal areas of concern are inaccessible by endoscopy.

Medical therapy remains the first line of treatment, especially for inflammatory strictures\textsuperscript{14,18}; however, endoscopic dilation of the affected area can also be utilized for strictures that are readily accessible, especially anastomotic strictures.\textsuperscript{73} The most commonly utilized approach is a “through-the-scope” method using 10- to 20-mm balloons, although a standardized methodology of sequential dilations has not been established.\textsuperscript{74} The typical indication is a limited, short-segment (<5 cm) stricture in the absence of associated penetrating disease (eg, abscess, fistula).

The most commonly reported complications following dilation are hemorrhage, perforation, and sepsis. Although the average technical success rate is nearly 90%\textsuperscript{75} the 5-year clinical recurrence rate is ~36%.\textsuperscript{76} Dilatation of strictures <4 cm in length in the setting of Crohn’s disease has been associated with the best long-term outcome.\textsuperscript{77} Patients usually require at least 2 dilations to achieve patency over a 5-year period.\textsuperscript{74} Experience is limited with other reported techniques including expandable metal or biodegradable stent placement,\textsuperscript{78} as well as the off-label use of intralesional glucocorticoid\textsuperscript{80} or infliximab injection,\textsuperscript{81} prohibiting a generalized recommendation until more experience is available.

2. Surgery is indicated for patients with symptomatic small-bowel or anastomotic strictures that are not amenable to medical therapy and/or dilation. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Surgery (eg, resection or strictureplasty) is warranted for small-bowel and anastomotic strictures when medical and/or endoscopic treatment fails to adequately improve the patient’s symptoms or when there is concern about concomitant malignancy (see below).\textsuperscript{82} Strictureplasty may be more appropriate for nonphlegmonous strictures in selected patients with diffuse involvement of the small bowel, existing or impending short-bowel syndrome, or rapidly recurring disease. The type of strictureplasty performed is largely based on the length of the stricture and the pliability of the affected bowel.\textsuperscript{83,84} Complications after strictureplasty have been reported in 4% to 15% of patients in large series, and include obstruction, bleeding, sepsis, perforation, and death.\textsuperscript{84,85} Older age,\textsuperscript{86} preoperative anemia,\textsuperscript{87} hypoalbuminemia,\textsuperscript{88} weight loss,\textsuperscript{89} emergency surgery,\textsuperscript{90} and the presence of an intra-abdominal abscess with peritoneal contamination\textsuperscript{90-92} are all reported risk factors for postoperative morbidity. Conversely, preoperative glucocorticoid use, number, site and length of strictureplasties, and synchronous bowel resection have not been associated with a higher rate of complications.\textsuperscript{85} Meta-regression analysis has reported 5-year recurrence rates for strictureplasty in jejunoileal and ileocolonic locations between 25% and 30%, including a ~3% site-specific recurrence, with the majority of recurrences developing in intestinal segments remote from the previous strictureplasty sites.\textsuperscript{85} Although patients undergoing strictureplasty alone are at risk for recurrence,\textsuperscript{93} this is, in general, comparable to resection.\textsuperscript{94} Factors such as malnutrition, the presence of phlegmon/perforation/fistula at the site, multiple strictures within a small segment, and suspicion of a malignancy are all relative contraindications to strictureplasty.\textsuperscript{94} The procedure can also be safely performed for disease involving the neo-terminal ileum associated with an ileocolostomy when short-bowel syndrome is a concern.\textsuperscript{95,96} However, the 5-year cumulative surgical recurrence rate in this location is ~45% and 24% for strictureplasty and resection of the ileocolostomy.\textsuperscript{97}

3. Patients with strictures of the colon that cannot be adequately surveyed endoscopically should be considered for resection. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Because of the transmural inflammation associated with Crohn’s disease, it is not uncommon to have luminal narrowing within the colon, especially with repeated flares. Obstructive symptoms from colonic strictures occur in up to 17% of patients.\textsuperscript{97} Even in the absence of symptoms, colonic strictures harbor occult carcinoma in approximately 7% of instances.\textsuperscript{97} It is often difficult to differentiate malignant from benign strictures on a strictly clinical basis. Short strictures, as opposed to long strictures, appear to be more commonly associated with cancer, as is advanced age and longer duration of disease (3.3% with less than 20 years of Crohn’s disease vs 11% with disease >20 years).\textsuperscript{97}
However, any colorectal stricture should be extensively assessed with multiple endoscopic biopsies to ensure the absence of malignancy. Cytological investigation of the large bowel (although not widely utilized) has been reported to aid in the diagnosis of colorectal carcinomas in this setting. Brushing with cytology appears to be an easy, highly specific, but relatively insensitive method to help in the diagnosis of colorectal cancer, and can be used as a complement to biopsy.86 If the stricture cannot be adequately surveyed to exclude a concomitant carcinoma, resection of the affected large intestine should be considered. In this situation, appropriate resection following standard oncological principles should be performed. Although colonic strictureplasty has been reported for segmental disease,89 it is generally discouraged owing to a lack of proven benefit, as well as concerns about concomitant carcinoma being left in situ.

Penetrating Disease

1. Patients with a free perforation should undergo surgery. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.

Patients with Crohn’s disease can present with free perforation of the small or large bowel, and perforation is the indication for surgery in ~1% to 16% of instances.100 Immediate resection of the perforated segment is preferred over simple suture closure101 because of a relatively high failure rate and concomitant increased mortality associated with the latter procedure.102 After resection of a small-bowel perforation, an anastomosis is typically possible. A diverting or an end stoma in the setting of a small-bowel perforation may be performed depending on the presentation and operative findings, and is often reserved for patients with severe hemodynamic instability, significantly edematous bowel, or technical challenges in constructing the anastomosis. In addition, a stoma following a small-bowel perforation may be needed in patients with other coexisting risk factors such as severe malnutrition or significant abdominal contamination. Benefits of stoma creation to divert a low-risk small-bowel anastomosis should always be balanced against the risk of additional surgeries that will subsequently be needed to close it.

In patients presenting with a free colonic perforation, a primary anastomosis with fecal diversion or an end stoma may be considered, especially in a patient presenting with feculent peritonitis, associated hemodynamic instability, moderate to severe protein-calorie malnutrition, chronic steroid or anti-TNF use, and treatment delay.103 Additional factors such as the extent of colonic resection, location of the anastomosis, baseline continence, and the presence or absence of proctitis or perianal disease also need to be taken into account when considering the need for diversion.104 Carefully selected patients (ie, hemodynamically stable, healthy bowel, technically sound anastomosis, minimal comorbidities) presenting with purulent peritonitis but without widespread feculent contamination may be suitable for a primary anastomosis. Finally, any unstable patient or those with severe peritoneal contamination from perforation at any site may be left in temporary discontinuity and brought back to the operating room for a washout and/or definitive procedure following appropriate resuscitation.

2. Patients with enteroparietal, interloop, intramesenteric, or retroperitoneal abscesses may be managed by antibiotics with or without percutaneous drainage. Surgical drainage with or without resection should be considered when this is not successful. Grade of Recommendation: Weak recommendation based on moderate-quality evidence, 2B.

Intra-abdominal abscesses in patients with Crohn’s disease typically result from a perforation or penetrating ulcers that are contained by surrounding structures, which may include adjacent segments of bowel. Small (<3 cm) abscesses can generally be sterilized with parenteral antibiotics alone.105 Antibiotics and percutaneous drainage (PD) of a larger abscess usually resolve symptoms of sepsis, infrequently results in an enterocutaneous fistula, and occasionally obviates the need for future surgery.106-111 Comparative analyses of PD and initial surgery for spontaneous abdominal abscess in Crohn’s disease indicate that successful PD, defined as abscess resolution and avoidance of subsequent surgery, occurs in the range of 23% to 78%.112-116 Factors associated with PD failure include concomitant steroid use, colonic disease, and abscesses that are large, multiloculated, or multifocal.115,117,118 In comparison with initial surgery, PD as a bridge to surgical resection results in a decrease in overall complications, diverting stomas, and overall cost, with similar rates of postoperative enterocutaneous fistula and anastomotic leak.115,116 Initial PD combined with intravenous antibiotics, steroids, total parenteral nutrition, and same admission bowel resection has been shown to result in the ability to perform a primary anastomosis (84%), with low risk of recurrent abscess and the need for subsequent surgery.119 The role of anti-TNF therapy in the setting of inflammatory mass (ie, phlegmon) with a concomitant abscess may be safe if the regimen includes initial intravenous antibiotics, followed by anti-TNF therapy, and continuation of antibiotics for 3 months total duration. With 2-year follow-up, it has been shown that most patients remain on anti-TNF therapy and do not require surgery.120 Following successful resolution of an abscess with PD, consideration should be given to performing a drain study to exclude a concomitant fistula before drain removal, although this practice has been largely anecdotal with no large-scale trials to evaluate its utility.

Although centers comparing surgical versus medical management of these abscesses report similar
Outcomes, efforts should be made to avoid surgery as the first line of treatment. When medical management is not successful, operative intervention with resection should be considered instead of operative drainage alone, the latter of which has been associated with higher rates of enterocutaneous fistula and reoperation. Overall, surgeons must make every effort to avoid overaggressive resection, because unnecessary removal of bowel can lead to immediate or future development of short-bowel syndrome.

3. Patients with enteric fistulas and symptoms or signs of localized or systemic sepsis that persist despite appropriate medical therapy should be considered for surgery. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

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 treated with appropriate resuscitation and parenteral antibiotics, and imaging studies should be performed to identify a source. If a concomitant abscess is identified, it should be drained when feasible. Regardless of whether an abscess is present or not, persistent sepsis usually warrants excision of the diseased bowel. Management of the affected organs is based on differentiating diseased bowel from other loops of bowel or organs that are secondarily involved and may or may not require resection. In general, diseased bowel is resected, whereas noninflamed bowel can be primarily closed, and other internal organs (e.g., bladder, vagina) are primarily closed or left to heal by secondary intention. Surgery may not be required in the absence of symptoms, because the presence of an internal fistula does not necessarily mandate surgery, especially in the absence of malabsorption, diarrhea, or recurrent infections (e.g., enterovesical fistulas).

Hemorrhage

1. Stable patients with significant GI hemorrhage may be evaluated and treated by endoscopic and/or interventional radiological techniques. Unstable patients should typically undergo operative exploration. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Acute lower GI hemorrhage is an unusual complication of Crohn’s disease, occurring in 0.9% to 6% of patients. Significant lower GI bleeding in patients with Crohn’s disease is more often secondary to severe inflammation and, unlike other sources of massive lower GI bleeding (e.g., diverticula, arteriovenous malformation), rarely necessitates the diagnostic evaluation required in a bleed of unknown origin. Patients with more significant bleeding should undergo concomitant resuscitation along with appropriate diagnostic measures. Stable patients can undergo CT imaging with prominent contrast arterial phase, endoscopy, and/or mesenteric arteriography. Although not specific to Crohn’s disease, CT angiography has reported sensitivity rates for acute GI bleeding of up to 89% and specificity rates of up to 92%. In addition, the latter 2 modalities also offer the benefit of allowing concomitant therapeutic intervention. The likelihood of identifying a precise bleeding source is somewhat limited, because spontaneous cessation of bleeding occurs in nearly one-half of all patients with lower GI bleeding. However, patients remain at risk for recurrent bleeding in nearly 40% of cases. More specific to patients with Crohn’s disease, the probability of rebleeding in those successfully managed with nonoperative measures may be reduced with anti-TNF agent therapy. Therefore, treating the underlying inflammation with the broad range of Crohn’s disease medications will often help minimize the risk of significant recurrent lower GI bleeding.

In general, surgical treatment is recommended in patients with life-threatening bleeding, persistent hemodynamic instability, or recurrent significant GI bleeding. If the source of bleeding has been localized by the use of preoperative or intraoperative modalities, a targeted resection with adherence to accepted surgical tenets is recommended. In the case of Crohn’s colitis, a total colectomy is recommended for patients who have persistent hemodynamic instability or persistent, serious bleeding that cannot be localized to 1 segment of the colon. The decision regarding creating an anastomosis versus an end or a diverting ileostomy should again depend on factors such as the patient’s degree of immunosuppression, age, hemodynamic stability, continence, and the presence of active Crohn’s disease in the rectum or perianal area.

Growth Retardation

1. Prepubertal patients with significant growth retardation despite appropriate medical therapy should be considered for surgery. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.

Crohn’s disease manifests itself during childhood or adolescence in up to 20% of patients, and the potential for linear growth impairment as a complication of chronic intestinal inflammation is unique to this population. Growth retardation exists in ~10% to 40% of children with Crohn’s disease, may present at the time of diagnosis or occur after diagnosis, and potentially causes compromised height at maturity. The treatment of young patients may include medical, nutritional, and surgical therapy that aims to decrease mucosal inflammation and alleviate symptoms, all while improving growth rates and promoting normal pubertal development. Resection of localized disease maximizes “catch-up” growth, and is warranted early in the disease process in otherwise
treatment-resistant, prepubertal children because they usually will achieve catch-up growth within the ensuing 6 months.144 However, early surgical intervention for growth retardation is only indicated in cases with clearly localized disease because of the risk for recrudescence with more diffuse involvement, as well as owing to the association between malnourishment and further developmental difficulties with extensive resections.

**Neoplasia**

1. **Patients with long-standing Crohn's disease of the ileocolic region or colon should have endoscopic surveillance of the large bowel. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.**

The incidence of colorectal carcinoma in a patient with Crohn's disease is 0.5/1000 person-years duration, which is a 2- to 3-fold increase in risk compared with the incidence in an age-matched general population.145 The impact of age at disease onset, disease duration, extent of disease, primary sclerosing cholangitis, and a positive family history of sporadic colorectal cancer are debated, but may be associated with an increased risk of colorectal cancer.146-150 Furthermore, postinflammatory pseudopolyps are thought to increase the risk of colorectal cancer, and inflammation is believed to be a risk factor for progression to colorectal neoplasia.151

All patients, regardless of the extent of disease at initial diagnosis, should undergo a screening colonoscopy no later than 8 years after the onset of symptoms with multiple biopsy specimens obtained throughout the entire colon to assess the true microscopic extent of inflammation.151,152 It has also been recommended that patients with more than one-third of the colon and rectum affected by Crohn's disease should be surveyed beginning within 1 to 2 years of the initial screening colonoscopy,151-153 and continued at 1- to 3-year intervals, despite a lack of high-quality evidence to support this recommendation.151,152 Surveillance colonoscopy should be ideally performed when the colonic disease is in remission. Although no prospective studies have determined the optimal number of biopsy specimens that should be acquired to reliably detect dysplasia, a minimum of 33 specimens has been recommended in patients with pancolitis.151,152,154 Chromoendoscopy with targeted biopsies is an alternative to random biopsies for endoscopists who have expertise with this technique, because limited reports have shown a higher sensitivity for detecting dysplasia by chromoendoscopy than with traditional white light endoscopy.155 Conversely, high-definition colonoscopy has been reported to improve targeted detection of dysplastic lesions during surveillance colonoscopy.156

Patients with concomitant primary sclerosing cholangitis (PSC) should undergo a screening colonoscopy when PSC is initially diagnosed, followed by annual surveillance colonoscopy with biopsies because of the reported increased risk of malignancy.151,152,157,158 The presence of concomitant PSC may also be a consideration for prophylactic total proctocolectomy, although the relative risk of malignancy for PSC patients and Crohn's is somewhat controversial, and appears to be less than that of PSC in patients with ulcerative colitis.159-161 Of note, when malignancy does occur in the setting of PSC and Crohn's disease, it has a predilection for occurring proximal to the splenic flexure.159 In addition, patients with a history of colorectal cancer in a first-degree relative, ongoing active endoscopic or histological inflammation, or anatomic abnormalities such as multiple inflammatory pseudopolyps or a stricture may also benefit from more frequent surveillance examinations or prophylactic bowel resection.

2. **Total proctocolectomy should be considered for patients with carcinoma, a nonadenoma-like dysplasia-associated lesion or mass (DALM), high-grade dysplasia, or multifocal low-grade dysplasia of the colon or rectum. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.**

Dysplasia is regarded to be the best indicator of colorectal cancer risk in Crohn's disease, although the diagnosis of dysplasia in mucosal biopsy specimens is associated with a high level of interobserver variability, especially when diagnosing indeterminate and low-grade dysplasia.162,163 Therefore, pathologists with expertise in GI diseases should normally confirm all cases diagnosed as indefinite, low-grade or high-grade dysplasia before a management plan is implemented, when possible.164

Unfortunately high-grade evidence detailing the natural history of dysplasia is still lacking, which ultimately affects surveillance and treatment recommendations. Although dysplasia is present in many patients with adenocarcinoma,165 colorectal cancer can also be the index neoplastic lesion166,167 or develop in the setting of low-grade dysplasia without intervening high-grade dysplasia.168 Furthermore, indefinite dysplasia and uniform low-grade dysplasia are associated with a higher incidence of synchronous and metachronous advanced neoplasms,166,167 although the magnitude and significance of that risk are still unknown.

A non-adenoma dysplasia-associated lesion or mass (DALM) is a raised, endoscopically visible lesion that harbors dysplastic mucosa and does not demonstrate the endoscopic feature of a sporadic adenoma. Nonadenomatous DALMs are an indication for resection because the incidence of associated cancer approaches 40%.166 Conversely, sporadic adenomas can develop in a background of Crohn's disease, and they can be safely managed by polypectomy and continued surveillance, especially if flat dysplasia is absent both from the mucosa immediately surrounding the polyp, as well as elsewhere in the colon.168,151,168
The extent of resection is influenced by the site of neoplasm and recognition that multifocal dysplasia is ultimately found in more than one-third of specimens from patients undergoing colectomy for low- or high-grade dysplasia detected during diagnostic or surveillance colonoscopy. Accordingly, resection of all large bowel (ie, total proctocolectomy) has been advocated over segmental resection. Further support for such an approach stems from observing that ~14% to 40% of the patients who undergo segmental resection for cancer of the large intestine develop metachronous colorectal cancers.  

3. Suspicious lesions (ie, mass, ulcer) identified in patients with Crohn’s disease should typically be biopsied, especially when considering a small-bowel strictureplasty. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Suspicious lesions anywhere along the GI tract in patients with Crohn’s disease warrant further evaluation. This requires a diagnosis confirmed by pathology when possible. The incidence of small-bowel carcinoma in a patient with Crohn’s disease is 0.3/1000 person-years duration, and this represents an 18.75-fold increase compared with an age-matched standard population. Although uncommon, a few cases of adenocarcinoma arising at or near a previous strictureplasty site have been reported. Biopsy of any suspicious mucosal lesions or suspicious ulcerations at the time of strictureplasty is accordingly suggested. Failure to do so may result in leaving malignant or premalignant lesions in situ that would otherwise warrant resection.

SITE-SPECIFIC OPERATIONS

Terminal Ileum, Ileocolon, and Upper GI 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. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.

Resection continues to be the most commonly performed operation for symptomatic inflammatory or penetrating disease affecting the small bowel and proximal colon, especially when existing or impending short-bowel syndrome is unlikely and medical management fails. Limited (2-cm) macroscopic disease-free resection margins are adequate, because this conserves bowel length and is not associated with an increased risk of disease recurrence. Certain factors are associated with an increased risk for infectious complications following surgery. In addition to steroids and immunomodulators, other associated factors include a concomitant abscess, or fistula, malnutrition (>10% weight loss within past 3 months), anemia (<10 g/dL), low serum albumin (<3.0 mg/dL), penetrating disease, prolonged operative times (>180 minutes), and urgent surgery. When one or more of these factors is present, surgery should be postponed, when possible, until the patient can be safely optimized. Alternatively, proximal fecal diversion may be more frequently indicated, although resolution of distal inflammation following diversion is more variable in the setting of IBD.

2. Select patients with symptomatic disease of the stomach or duodenum should be considered for endoscopic dilation, bypass, or strictureplasty of the affected area. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Gastroduodenal disease occurs in 0.5% to 4% of all patients with Crohn’s disease. Obstructive symptoms arising from single, short, moderately thick strictures of the duodenum can be successfully treated with endoscopic dilation as an initial procedure. Symptomatic Crohn’s disease of the stomach or duodenum requiring operative treatment is often best managed by bypass (ie, gastrojejunostomy, duodenojejunostomy) or strictureplasty, rather than resection. Proton pump inhibitor therapy has supplanted the need for concomitant vagotomy in patients at risk of sustaining marginal or duodenal ulceration following bypass. Because the operation has become more simplified, a laparoscopic approach may be considered because of its decreased risk for operative morbidity and equivalent recurrence rates. Nonperforated, nonphlegmonous stenotic lesions in this region can alternatively be managed by strictureplasty. Finally, solitary strictures of the second and third portions of the duodenum are often best suited for strictureplasty.

Colon

1. The procedure of choice for emergency surgery in Crohn’s colitis is a total abdominal colectomy with end ileostomy. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.

Subtotal or total colectomy with construction of an end ileostomy and Hartmann closure of the distal bowel or creation of a mucous fistula is a safe procedure in patients with severe or fulminating Crohn’s colitis who require surgery. This approach removes most of the inflamed intestine with a relatively simple operation that avoids a pelvic dissection and the risks of an anastomosis in this setting. Extrrafascial placement of the closed rectosigmoid stump may be associated with fewer pelvic septic complications than an intraperitoneal position, although it may be technically difficult to achieve. Abdominal drains as well as transanal drainage of the distal stump may further decrease the risk of pelvic sepsis.
2. Patients with colonic disease and rectal sparing that require elective surgery may undergo segmental colectomy for single-segment disease or total colectomy for more extensive disease. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.

Outside the aforementioned concerns regarding malignancy, symptomatic disease of the colon with rectal sparing can be managed by removal of only the diseased segment or by total abdominal colectomy. The operations are comparable with respect to operative complications, need for a permanent stoma, and risk of recurrence; however, segmental resection is associated with surgical recurrence that occurs a median of 4.4 years earlier. Total colectomy with ileoproctostomy is preferable if 2 or more colonic segments are affected, because this subgroup has a higher recurrence rate when segmental resections are performed.

3. Patients who require surgery for disease of the rectum typically undergo total proctocolectomy, or proctectomy with creation of a stoma. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Refractory proctitis generally warrants total proctocolectomy with a permanent ileostomy owing to a frequent association with colonic involvement; however, proctectomy alone with creation of a colostomy can be considered if the colon is spared. When proctectomy is warranted, the entire rectum should be resected because carcinoma has been described in patients with even a short Hartmann remnant. Furthermore, an intersphincteric dissection and primary closure of the perineal wound is preferred, because it is associated with fewer wound complications and less morbidity compared with a traditional abdominoperineal resection.

4. Total proctocolectomy with IPAA is not generally recommended for patients with known Crohn’s disease. Grade of Recommendation: Strong recommendation based on low- or very low-quality evidence, 1C.

Two centers have suggested that a restorative proctocolectomy with an IPAA can be considered in Crohn’s disease in highly selected patients if the small bowel is unaffected and there is no history of perianal disease. In appropriately selected patients, the 5-year pouch survival rate is at least 85%. However, this must be balanced against the high risk of further complications, pouch loss, poor function, and the need for further immunosuppressive therapy. Therefore, although there are advocates for restorative proctocolectomy in specific cohorts, this should be approached with extreme caution, and at the present time, is not widely recommended.

TECHNICAL CONSIDERATIONS

1. Patients who require resection of the affected bowel should be offered a laparoscopic approach where appropriate experience and expertise are available. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.

A laparoscopic approach is preferred to an open operation where appropriate surgeon expertise is available. For small-bowel and ileocolic Crohn’s disease, laparoscopy has been associated with earlier return of bowel function, shorter length of stay, and fewer postoperative complications. Long-term outcomes, including clinical recurrence, do not appear to be compromised with a laparoscopic approach. Furthermore, a laparoscopic approach can also be safely used when managing complex (eg, penetrating, recurrent) disease, when appropriate surgical experience and expertise is available. Single-port laparoscopy may be also considered by those facile with this approach, but its advantages over multiport laparoscopy have not been consistently described.

When performing a total abdominal colectomy for Crohn’s colitis in the elective setting, laparoscopy is associated with decreased perioperative morbidity and improved short-term outcomes (ie, length of stay, return of bowel function) in comparison with an open approach. The risk of wound infection and intra-abdominal abscess are significantly lower with a laparoscopic total abdominal colectomy compared with open surgery in the patient with nontoxic Crohn’s disease. Even in an emergent setting, a laparoscopic approach for severe acute Crohn’s colitis that failed adequate medical management is associated with a shorter length of stay and fewer minor morbidities.

2. Patients who are candidates for primary anastomosis following resection of diseased intestine can have the anastomosis constructed as deemed most appropriate by the surgeon. Grade of Recommendation: Strong recommendation based on moderate-quality evidence, 1B.

Despite earlier reports, the anastomosis following resection in the setting of Crohn’s disease can be configured in any manner (eg, end-to-end, side-to-side, end-to-side) without increasing the risk for endoscopic, clinical, or operative recurrence. A side-to-side (ie, functional end-to-end) anastomosis constructed with staples or sutures does not reduce the likelihood for anastomotic leak compared with other configurations and techniques. However, it may reduce the overall rate of postoperative complications. Although a stapled side-to-side anastomosis has been shown to reduce reoperation rates, the use of the mechanical stapler may be contraindicated when the bowel wall is thickened as seen with bowel proximal to a chronic obstruction.
APPENDIX A

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REFERENCES


