Crohn's Disease of the Colon, Rectum, and Anus

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KEYWORDS

• Crohn's Disease • Perianal Crohn's Disease • Colonic Crohn's Disease
• Crohn fistula • Ileostomy

KEY POINTS

- Intra-abdominal abscesses associated with colonic Crohn's Disease (CD) that are addressed before definitive surgery may be associated with fewer stomas.
- Segmental colectomy for colonic CD is a viable option for patients with limited disease.
- The definitive operation for the patient with colonic CD is total proctocolectomy with end ileostomy (TPC/I).
- Fecal diversion when done to decrease colonic inflammation, perianal inflammation, and sepsis may become permanent.

CD of the large intestine is one of the more challenging forms of the disease to treat.[1] CD of the small bowel and ileocolic disease are often treated surgically without the need for an ostomy, whereas the decision tree for surgical treatment of CD of the colon, rectum, and anus frequently has an ostomy at an early branching point. Also, because an ostomy has such a life-changing impact, CD of the large intestine can require difficult decisions. Delays in treatment because of the fear of an ostomy can also lead to more complicated procedures. This review focuses on the less-common complications of CD of the colon, rectum, and anus as well as the surgical treatment options.

PRESENTATIONS OF CROHN'S DISEASE

Intra-Abdominal Abscess

Intra-abdominal abscesses can complicate the treatment of patients with CD and add additional steps in management. These steps can include percutaneous drainage, surgical drainage, and/or fecal diversion. Ideally, preoperative drainage of an abscess would obviate surgery in the acute setting, make future surgery technically easier for the surgeon and patient, and decrease the likelihood of the need for an ostomy.

The author has no conflicts of interest.
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Surg Clin N Am 95 (2015) 1195–1210
http://dx.doi.org/10.1016/j.suc.2015.07.005 **surgical.theclinics.com**
0039-6109/15/$ – see front matter

da Luz and colleagues[2] retrospectively reviewed the Cleveland Clinic, Ohio, experience with abdominal and pelvic abscesses in patients with CD from 1997 to 2007 and evaluated the efficacy of percutaneous drainage of abscesses in 94 patients. Patients with postoperative and perirectal abscesses were excluded from this review. Of this group of patients, 82% had ileocolic CD and 16% had colonic CD. An abscess was the initial presentation of CD in only 5 patients; 31 of 48 patients (65%) had what was considered a successful delay in surgery (median delay of surgery of 43 days). Factors associated with failure of percutaneous drainage were steroid use, colonic disease phenotype, and multiloculated abscesses. However, the size of the abscess was not associated with success or failure. As should be the goal with preoperative percutaneous drainage of intra-abdominal abscesses, initial percutaneous drainage did reduce the incidence of stoma creation when compared with initial surgical intervention (23% vs 58%, $P = .01$).

The nonsurgical viewpoint of intra-abdominal abscesses was summarized by an article from Massachusetts General Hospital.[3] Gutierrez and colleagues[3] reviewed 66 patients who were treated for intra-abdominal abscesses from 1991 to 2001. Of these, 37 patients had percutaneous drainage and 29 patients had surgical drainage of abscesses. The evaluation focused on the time to resolution of abscesses, which was not different between percutaneous and surgical drainage. The investigators also evaluated the need for surgery after percutaneous drainage and found that one-third of patients required surgery within 1 year. The investigators did not comment on whether or not patients treated with percutaneous drainage had a lower incidence of stoma creation.

Abdominal Wall Abscesses

In contrast to intra-abdominal abscesses, abdominal wall abscesses are less common. Abscesses of the abdominal wall typically indicate an underlying fistula. Neufeld and colleagues[4] identified 13 patients over a 10-year period who were diagnosed with abdominal wall abscesses resulting from CD. Mean patient age was 32.8 years. In 2 patients, the abscess was the initial presentation of CD; 6 patients were found to have colonic CD, and 5 patients had ileocecal CD. All 13 patients ultimately had definitive surgery with resection of the source of the fistula. As the fistula is the source of the abscess, it must be addressed. The investigators noted that draining the abdominal wall abscess without addressing the fistula led to a 100% failure rate, and this is in contrast to percutaneous drainage of intra-abdominal abscesses that are not associated with fistulae. It was concluded that the presence of an abdominal wall abscess indicates complicated CD and that preoperative drainage can prepare the patient for definitive surgery.[4]

Unfortunately, much of the literature describing abdominal wall abscesses does not differentiate intra-abdominal abscesses from retroperitoneal (psoas) abscesses. A psoas abscess is most commonly the result of a mesenteric abscess extending through the mesentery into the retroperitoneal space overlying the psoas muscle.[4] The formerly (now rare) classic presentation of an iliopsoas abscess is septic arthritis of the hip.[5] While this was previously most commonly seen with spondylitis resulting from tuberculosis, CD is now a much more common cause.[4]

Toxic Megacolon

Toxic megacolon is uncommon in both ulcerative colitis (UC) and CD, occurring in only 1% to 5% of patients with CD. This condition may occur as an exacerbation of inflammatory bowel disease (IBD), but it is the initial manifestation of IBD in more than 60% of patients.[6] Multiple factors have been shown to precipitate toxic megacolon, including antidiarrheal agents, belladonna alkaloids, and opiates.[7] The criteria for toxic

megacolon are not only dilation of the colon but also systemic symptoms of toxicity. Jalan and colleagues[8] proposed criteria for the diagnosis of toxic megacolon in 1969. These criteria, which are still commonly used, include a colonic diameter of 6 cm or greater and 3 of the following:

1. Temperature greater than 38 °C
2. Heart rate greater than 120 beats per minute
3. Anemia
4. White blood cell count greater than 10.5×10^9/L.

The criteria also require that one of the following conditions be present:

1. Hypotension
2. Altered mental status
3. Dehydration
4. Electrolyte abnormalities[9]

Treatment of toxic megacolon includes broad-spectrum antibiotics as well as corticosteroids. Early surgical consultation is recommended.[8] If improvement does not occur within 24 to 72 hours, surgery is recommended; TAC with end ileostomy is the procedure of choice.[7] Early surgical intervention may prevent colonic perforation. Colonic perforation that occurs with toxic megacolon is associated with a mortality rate of 20%, when compared with a 4% mortality associated with toxic megacolon without perforation.[10] Unlike other cases of megacolon such as colonic pseudo-obstruction (Ogilvie syndrome), colonoscopy is generally contraindicated in cases of toxic megacolon. *Clostridium difficile* colitis, as well as colitis resulting from cytomegalovirus infection should be excluded.[10,11]

Bleeding

Massive gastrointestinal (GI) tract bleeding may occur in the setting of CD and can be particularly difficult to treat. Typical diagnostic procedures that are used in patients with lower GI tract bleeding without IBD are often also used in patients with bleeding resulting from CD. These procedures include colonoscopy and angiography. Kostka and Lukas[12] identified 6 of 156 patients with CD over an 18-year span who had massive GI tract bleeding; 3 patients had a previous diagnosis of CD, whereas 3 patients presented with bleeding as the initial symptom of CD. These 6 patients had a total of 11 episodes of bleeding. The site of bleeding was identified preoperatively by angiography in 2 patients and by colonoscopy in 2 patients. The most common procedure required was an ileocecal resection (3 patients). Of the 6 patients, 4 ultimately underwent surgery, which led the investigators to conclude that a conservative approach may be tried as first-line therapy, but surgery is inevitable.

Robert and colleagues[13] reviewed the Mount Sinai, New York, experience with massive GI tract bleeding resulting from CD. At a center with an extensive experience with IBD, over a 26-year period (1960–1986), only 21 of 1526 patients developed severe GI tract bleeding. The group of 11 men and 10 women had a median age of 27 years. A total of 12 surgical procedures were done including 6 subtotal colectomies, 4 ileocolonic resections, 1 sigmoid colectomy, and 1 right hemicolectomy. Interestingly, 9 of the 10 patients with ileocolonic CD also had colonic involvement. The frequency of bleeding was much higher among patients with colonic involvement (17/929; 1.9%) when compared with patients with small bowel disease (4/597; 0.7%) ($P<.001$). Recurrent bleeding was quite unusual.[13] Although the differences were not statistically significant, they commented that surgery at the time of the initial bleeding seemed to lead to a lower mortality and a lower chance of recurrent bleeding.

Several investigators have stated that ileocecal CD was the most common form of CD that caused bleeding, whereas Belaiche and colleagues[14] found the opposite. These investigators retrospectively identified 34 cases of CD with GI tract bleeding. The site of bleeding was attributed to the colon in 85% of cases when compared with isolated small bowel disease in only 15% ($P<.0001$). An ulcer was identified to be the cause of the bleeding in 95% of patients, and this ulcer was most commonly in the descending colon.

However, not all investigators think that surgical resection is the only treatment option. A recent retrospective review from Thailand identified 7 patients who were not considered fit for surgery (3 were poor surgical candidates, and 6 were thought to be at high risk of short bowel syndrome). Of the 7 patients, 5 presented with bleeding as their first presentation of CD. These patients did not undergo angiographic embolization because of the risk of short bowel syndrome. These patients were treated with infliximab (5 mg/kg). Bleeding stopped within 24 hours in 6 of 7 patients, and 1 patient had an episode of rebleeding, which stopped 10 days after the initial infusion of infliximab. The median number of doses of infliximab was 2.[15]

Obstruction Because of Stricture

Strictures do occur in the large bowel in patients with CD, but the frequency is less than that in the small bowel. Colonic strictures are more common in patients with UC than in those with CD, occurring in approximately 5% to 17% of patients with UC and 5% of patients with CD.[16] Although many would think that colonic strictures are more likely to be malignant in patients with CD than in those with UC, the converse has actually been shown. Gumaste and colleagues[17] from Mount Sinai Hospital in New York found 29% incidence of malignancy in patients with UC who had colonic strictures, when compared with a 6.8% incidence of malignancy in those with Crohn strictures.[18]

Strictures may be classified as either inflammatory or fibrostenotic lesions. Differentiating between the 2 is important for determining appropriate treatment.[19] The type of stricture may be differentiated based on imaging studies. Characteristics such as edema, bowel wall thickness, and vascularization help make this differentiation. The typical location of obstruction and strictures in CD is ileocecal.

It should be recalled that any stricture found in the colon or rectum should be biopsied because these should be considered malignant until proven otherwise. The role of strictureplasty in patients with colonic strictures has yet to be defined, but many consider it contraindicated.[16]

OPERATIONS AND TECHNIQUES FOR CROHN'S DISEASE OF THE COLON AND RECTUM

Multiple different surgical operations exist for the patient with CD of the colon and rectum. Several pieces of information should be taken into consideration when deciding which operation is most ideally suited for the individual patient. The amount of colon involved with the colitis should be considered, as well as whether or not the rectum is involved. A digital examination of the anus and distal rectum should be done because perianal CD may contraindicate a sphincter-saving operation and cause the surgeon to recommend an operation with an ostomy. In addition, the function of the anus should be considered, and any underlying fecal incontinence may lead to a change in procedure.

Segmental Resection

A common dilemma for the surgeon faced with a patient with colonic CD and rectal sparing is whether the patient should have a segmental resection or a TAC. Martel

and colleagues[20] reviewed their experience with segmental colectomy for colonic CD. A total of 84 patients underwent segmental colectomy, 32% patients required an ostomy, and 36 patients underwent reoperation. The mean time to reoperation was 4.5 years. The investigators concluded that there was no higher risk of surgical recurrence in patients undergoing segmental colectomy when compared with those undergoing TAC/IRA.

Fichera and colleagues[21] reviewed their outcomes of patients with Crohn colitis undergoing surgery. Over an 18-year period, 179 patients with colonic CD had surgery, and 54 patients (30%) had a segmental colectomy done. Patients who had a segmental colectomy done had a statistically significantly higher risk of surgical recurrence than those who underwent TAC/IRA or TPC/I. Significantly, after 1 year, 30 patients who had segmental colectomy (61%) were still taking steroids or immunomodulators; 17 patients required a permanent stoma (34.7%). There was a statistically significant higher chance of the need for a stoma in patients with distal disease when compared with those with proximal disease. The investigators concluded that patients with involvement of multiple colonic segments, especially if the disease was distal (descending and rectosigmoid colon), had a much lower risk of recurrence if treated with TPC/I rather than segmental colectomy. However, they also noted that if patients have short-segment colonic CD (<20 cm), then segmental colectomy is a valuable alternative.[21]

Tekkis and colleagues[22] published a meta-analysis comparing TAC/IRA to segmental colectomy. Six studies from 1988 to 2002, which included 488 patients, were included. There was no difference in recurrence of CD between those undergoing TAC/IRA and those undergoing segmental colectomy. However, patients who had segmental colectomy did have recurrence of their disease a mean of 4.43 years earlier than patients undergoing TAC/IRA. It was concluded that segmental colectomy was an attractive option for select patients with colonic CD. However, in patients with colonic involvement of 2 or more segments, TAC/IRA was the preferred alternative.

Total Abdominal Colectomy with Ileorectal Anastomosis

An option for the patient with colonic CD without rectal disease (rectal sparing) or perianal disease is a TAC/IRA. The surgeon must ensure that not only the rectum but also the anus is free of inflammation. Although this will not be an option for all patients with CD of the colon and rectum, approximately 25% to 50% of patients will not have disease of both the rectum and anus and would therefore be candidates. The surgeon should exclude any other conditions, such as fecal incontinence, that would preclude an anastomosis.[23]

Fortunately, recurrence (or the new occurrence) of CD in the rectum or anus after TAC/IRA is uncommon. Longo and colleagues[23] reviewed 131 patients who underwent TAC/IRA. It should be noted that their statistical methods only included crude fractions and not a log rank test, such as Kaplan-Meier. Of the 118 patients with a functioning anastomosis, 30 patients required proctectomy and 16 patients required diversion with an ostomy. At a mean follow-up of 9.5 years, 72 patients retained a functional anastomosis. The mean bowel frequency was 4.7 per day.

O'Riordan and colleagues[24] also studied the long-term outcome of the IRA after TAC/IRA. Over a 28-year period, 81 patients underwent TAC/IRA for CD. The anastomotic leak rate was 7.4%. At 5 years, 87% of patients (95% confidence interval [CI], 75.5–93.3) had a functioning IRA, and at 10 years, 72.2% of patients had a functioning anastomosis (95% CI, 55.8–83.4). The investigators concluded that TAC/IRA is a good option for select patients but that the long-term risk of proctectomy is approximately 30%.

Restorative Proctocolectomy

While restorative proctocolectomy (proctocolectomy with ileal j-pouch-anal anastomosis [IPAA]) is considered by many to be the gold-standard operation for UC,[25] its use in patients with CD is controversial and much less common. In fact, many think that IPAA is contraindicated in patients with a preoperative diagnosis of CD.[26] The literature regarding IPAA is somewhat difficult to interpret because many patients with CD who undergo IPAA have a preoperative diagnosis of indeterminate colitis or UC and are only diagnosed with CD after surgery.[27] For those surgeons who do offer restorative proctocolectomy to patients with CD, preoperative requirements include the absence of both perianal and small bowel disease. Panis and colleagues[28] were some of the initial advocates for IPAA in the setting of CD because they began offering IPAA to patients with CD in 1985. Between 1985 and 1992, 31 patients with a preoperative diagnosis of CD underwent IPAA. Only 2 patients developed fistulae and 2 patients developed recurrent CD. No patients were lost to follow-up, and there was no statistically significant difference in stool frequency when their patients with CD were compared with 44 patients who had UC ($P = .68$).

Regimbeau and colleagues[29] have updated their experience with IPAA for CD. Although 26 patients had a preoperative diagnosis of CD, these results are still difficult to interpret because they are combined with the results of 15 patients who had CD diagnosed after IPAA was done. It is notable that 27% of the patients (11 of 41) had postoperative complications, including 7 patients who developed pouch-perineal fistulas. In addition, 38% of patients developed pouchitis and 3 patients (7% of the 41 patients) ultimately required pouch excision.[29]

The results of Regimbeau and colleagues[29] are in contrast to those of Sagar and colleagues[27] from the Mayo Clinic. These investigators reviewed 37 patients who underwent IPAA and were diagnosed postoperatively with CD. Of these 37 patients, 11 developed complex fistulas. Pouch failure occurred in 17 patients. The investigators concluded that IPAA done with a preoperative diagnosis of CD had a high failure rate (45%) but that long-term functional results were acceptable if the pouch was able to be preserved. Others have seen similar results to those of Sagar and colleagues.[27] Mylonakis and colleagues[30] identified 23 patients who had IPAA for CD. Of these patients, 48% had pouch excision after a mean follow-up of 10.2 years. Although IPAA for CD does have its advocates, CD remains an absolute contraindication to IPAA in the opinion of many surgeons.[21]

Total Abdominal Colectomy with Ileostomy Without Proctectomy

TAC with end ileostomy while leaving the rectum in situ (TAC/I) is an option for the patient with colonic CD in whom the pelvic dissection necessary for removal of the rectum is not thought to be wise. This option is an ideal one for a patient with an acute presentation of colonic CD requiring surgery (such as a patient with toxic megacolon or bleeding from a colonic source) or a patient with a more chronic disease presentation requiring surgery (ie, a patient on high doses of steroids).

Management of the Rectal Stump

When colectomy without proctectomy is done for an acute presentation of CD, there are different choices regarding how to deal with the rectal stump. One option is to merely close off the stump.[31] When this is done, drains should be placed in the pelvis; consideration should be given to placing a rectal tube for drainage as well. Some have noted that there is a chance of the stump blowing out when this option is chosen. In order to decrease the incidence of this event, some have placed the rectal stump in

the subcutaneous tissue of the abdominal incision with the hopes that if this does occur, it will occur in the wound rather than intra-abdominally. This placement would hopefully avoid an intra-abdominal or pelvic abscess. Others have noted that it is difficult to make the rectal stump reach the abdominal wall, and they have placed abdominal/pelvic drains, as well as a rectal tube, to lessen the chance of this occurring.[32] Trickett and colleagues[33] reported the outcomes of 37 patients who had an emergency TAC. In 27 patients, the stump was left in an intraperitoneal position, and it was placed in a subcutaneous position in 10 patients. Two patients had leakage from an intraperitoneal stump, which did not require further surgery but did prolong their length of hospital stay; 3 patients had leakage from a subcutaneously placed stump, which resulted in a mucous fistula and a wound infection. However, the subcutaneous placement of the rectal stump and the resulting wound infection did not seem to prolong the length of the hospital stay. The incidence of pelvic sepsis seems to be related to the length of the remaining rectum. The highest incidence seems to be with a short intrapelvic stump (33%), followed by an intraperitoneal stump (6%–12%), with a subcutaneously placed stump having the lowest risk of pelvic sepsis (3%–4%).[33] While one advantage of a longer stump is a lower incidence of pelvic sepsis, another advantage is that any future pelvic surgery is technically easier because it should be easier to dissect a longer stump. For example, in some situations, one may not be able to differentiate between CD and UC before surgery and a restorative proctocolectomy may be planned at some point in the future for the patient with UC.

Secondary Proctectomy

When the rectum is not removed at the time of the initial operation, secondary proctectomy may be required at a later time for Crohn proctitis or diversion proctitis. Common symptoms of diversion proctitis that develop after colectomy with an ostomy, such as mucous per anus and rectal bleeding, may become symptomatic enough to require treatment. Although Crohn proctitis does not usually require treatment after colectomy with an ostomy, diversion proctitis that develops may require treatment in a minority of patients.[34] If such treatment (commonly topical therapy) fails, secondary proctectomy may be required.

Cattan and colleagues[35] retrospectively reviewed 144 patients who underwent TAC for CD over a 17-year period. Of these, 118 patients had an IRA (the majority were not done at the time of the initial surgery, but were done a median of 3 months after surgery). Most patients who did not have an IRA done, had proctitis (the minority had perianal disease only or both proctitis and perianal disease). Thirteen patients underwent a secondary proctectomy, which was most commonly done for intractable proctitis (10 patients).

Geoghegan and colleagues[36] have also reviewed the need for secondary proctectomy in patients with CD who underwent TAC. A total of 44 patients had surgery for colonic CD. Secondary proctectomy was done in 5 of 12 patients who had surgery for acute colitis, but only in 1 patient who had an elective colectomy. None of their patients developed cancer in the retained rectal stump.

Rieger and colleagues[37] reviewed 38 patients who had TAC for CD between 1968 and 1994. Fourteen patients ultimately underwent secondary proctectomy (9 of these 14 patients had secondary proctectomy after TAC/I and 5 required proctectomy after TAC/IRA). The reader would be wise to recall that the experiences of both Rieger and colleagues[37] and Geoghegan and colleagues[36] was reported before the era of biological therapy for IBD, hence the requirement for secondary proctectomy is most likely lower now after the initiation of this therapy.

Additional reasons for proctectomy after TAC/I are the development of cancer or the inability to exclude cancer, as well as persistently symptomatic perianal CD. The development of cancer in the retained rectum after TAC/I is rare.[38]

Total Proctocolectomy with Ileostomy

The definitive operation for CD involving the colon and rectum is the removal of the entire colon and rectum with a permanent ileostomy. This operation has the advantage of removing the diseased intestine in one operation without a frequent need for secondary operations. Therefore, diseases and complications related to the rectum and rectal stump are not a concern. Recurrence of CD in patients undergoing TPC/I is much less than that in patients undergoing operations in which an anastomosis is done (such as segmental colectomy or TAC/IRA). Yamamoto and colleagues[39] reviewed their experience with TPC/I in 103 patients who had surgery between 1958 and 1997. The 5-, 10-, and 15-year recurrence rates were 13%, 17%, and 25%, respectively, after a median follow-up of 18.6 years. The investigators concluded that TPC/I carried a low long-term recurrence rate but that young males were at an increased risk of recurrence. Fichera and colleagues[21] reviewed 179 patients who had surgery for colonic CD. A total of 75 patients had TPC/I, and only 4 of these patients developed recurrent disease. The investigators noted that patients undergoing TPC/I were less likely to be taking medications 1 year after surgery than patients who had either segmental colectomy or TAC/IRA. They concluded that TPC/I was associated with low morbidity, longer time to recurrence, and a lower overall risk of recurrence.

Abdominoperineal Resection for Distal Crohn's Disease

For the patient with CD of the rectum or perineal CD, when the proximal colon is normal, an option is removal of the rectum and anus only with an intersphincteric proctectomy. de Buck van Overstraeten and colleagues[40] reviewed 10 patients who underwent abdominoperineal resection (APR) for CD. The proximal colon was found to be normal at colonoscopy. The investigators identified recurrent disease in 9 of 10 patients at a median of 9.5 months. Even with medical treatment, completion colectomy was done in 5 of these 9 patients. It was concluded that even with a normal-appearing proximal colon, TPC/I was a more appropriate procedure than APR in patients with Crohn proctitis and perineal disease because of the severe and early recurrence of CD.

MANAGEMENT OF SPECIFIC COMPLICATIONS AFTER SURGERY FOR CROHN'S DISEASE

Perineal Wound after Proctectomy

The perineal wound that is created after proctectomy for CD does not always heal primarily, and delayed healing after proctectomy has been a common difficulty. This fact is especially true in the patient who has perineal CD.[41] In some patients, this can be averted. The patient with an anorectal abscess should have this drained before definitive proctectomy. In addition, some patients with severe perianal CD with abscesses and fistulae may benefit from fecal diversion before definitive proctectomy. This procedure may increase the chance of primary healing and decrease the incidence of the nonhealed perineal wound. Yamamoto and colleagues[39] retrospectively reviewed 145 patients who underwent proctocolectomy for CD from 1970 to 1997; 33 patients (23%) developed a persistent perineal sinus after TPC/I. Multivariate analysis showed age, rectal involvement of CD, and fecal contamination at the time of surgery as risk factors for persistent perineal sinus. The investigators also noted that an extrasphincteric dissection was an additional risk factor for a persistent perineal sinus.

High Ileostomy Output after Total Proctocolectomy with End Ileostomy

High ileostomy output, known also as ileostomy diarrhea, can be quite disabling for some patients with CD who have had TPC/I. Some patients seem prone to this complication, such as those who have had a previous cholecystectomy (bile-acid-induced diarrhea) or the patient who has had a prior ileal resection for CD. In addition, dehydration is common when output is greater than 2 L/d (normal ileostomy output is 800–1000 mL/d).[42] Other common complaints in the patient with dehydration from high stoma output include decreased urine output and concentrated-appearing urine.[42]

Treatment should begin with the restriction of hypotonic fluids. The luminal concentration of sodium must be increased in order to cause sodium to be reabsorbed. Sodium absorption in the jejunum is coupled with glucose absorption; therefore, oral intake of a glucose-saline solution will lead to increased sodium reabsorption and a concomitant decrease in ileostomy output.[42]

Initial medical therapy should begin with antimotility agents, such as loperamide. Loperamide has the advantages of being an over-the-counter product and being nonaddictive. Other medical treatment options for patients with continued high ileostomy output include codeine phosphate, Lomotil (diphenoxylate hydrochloride and atropine sulfate), and tincture of opium. The use of the long-acting somatostatin analog octreotide has been reported in patients with high stoma output.[43]

While treatment of ileostomy diarrhea is usually successful with loperamide, other more novel treatment regimens have been studied. One novel treatment option for ileostomy diarrhea is the use of budesonide. Ecker and colleagues[44] studied 23 patients with CD and high ileostomy output (defined as >1000 mL/d). No other antiinflammatory agents were allowed. The patients were treated with budesonide 3 mg orally thrice daily for 4 weeks. Preoperative evaluation revealed no evidence of active inflammation. Budesonide was withdrawn, and there was a statistically significant increase in ileostomy output of 295 g/d; 20 of 22 patients had an increase in ileostomy output. Budesonide was reintroduced, and ileostomy output decreased by 323.7 g/d. Twenty patients had a decrease in ileostomy output. The investigators concluded that the antidiarrheal effect of budesonide is independent of its antiinflammatory effect.[44]

Perianal Crohn's Disease

Although CD was first described as a unique entity in 1932,[45] perianal disease was first described in 1959.[46] Perianal Crohn's Disease (PCD) is more commonly found in patients with CD involving both the colon and rectum.[47] Hellers and colleagues[48] found that 92% of patients with CD involving the colon and rectum had perianal disease, whereas only 41% of patients with CD with rectal sparing had perianal involvement. PCD is much less common in patients with small bowel or ileocolic CD, as Hellers and colleagues[48] found that patients with isolated ileal disease had an incidence of perianal disease of 12%, whereas 15% of patients with ileocolic CD had perianal disease.

Anorectal findings in PCD include not only anal fistulae but also anal tags, anal fissures, hemorrhoids, perianal abscesses, and strictures. Michelassi and colleagues[49] reported on 224 patients with anorectal complications of CD; 66 patients had a combination of anorectal findings. Other common findings included anal fistulae (51 patients) and anorectal abscesses (36 patients). Forty patients had anal stenosis.[49]

Besides anatomic findings, PCD can also be classified by the activity and impact on quality of life. A scoring system for gauging the activity of PCD has been developed.[50] The Perianal Disease Activity Index takes into account fistula activity, restriction of

activities, and the severity of perianal disease. The score is based on a 5-point scale and has been validated. The status of the rectum is also an important consideration in selecting treatment options because active proctitis resulting from CD will have a negative impact on perianal disease as well as decrease the number of possible treatment options for PCD.[51]

Treatment options for PCD depend on the type of findings. The anatomy of disease in CD is clearly different from that of a typical cryptoglandular disease, and therefore, the chance of healing after surgical treatment is also different. For patients with significant disease, fecal diversion in the form of an ostomy may be required.

Anal Fistulae

For patients with anal fistulae, several treatment options exist. Patients with simple (subcutaneous or intersphincteric) and singular anal fistulae may be treated with fistulotomy. However, in the patient with CD and multiple or complex anal fistulae, non–sphincter-preserving procedures such as fistulotomy should be used sparingly because of the risk of fecal incontinence.[51] Therefore, when a patient with PCD has multiple fistulae or fistulae that involve a significant amount of anal sphincter, consideration should be given to sphincter preservation. These patients are best treated with draining setons while control of their CD is obtained. As will be discussed, this may require medical therapy, and occasionally, fecal diversion. After this has been achieved, more definitive treatment of the fistulae can be initiated. This treatment may include anal fistula plugs, rectal advancement flaps, ligation of the intersphincteric fistula tract (LIFT procedure), or an anal fistulotomy.

Anal fistula plugs have also been used in patients with PCD. However, the use of such plugs has been studied much less extensively in patients with PCD than in those with cryptoglandular anal fistulae. In fact, several of the randomized trials studying anal fistula plugs have specifically excluded patients with CD.[52] A recent systematic review of the use of anal fistula plugs evaluated the efficacy in patients with fistulae related to CD.[53] Although the efficacy for anal fistula plugs was similar between patients with cryptoglandular anal fistulae and fistulae related to CD (55% vs 54%), the investigators concluded that the anal fistula plug had not been adequately studied in patients with CD.

Rectal mucosal advancement flaps have also been studied in PCD. A recent systematic review of endorectal advancement flap revealed that flaps done for cryptoglandular disease were much more successful than those done for CD (81% success rate vs 64% success rate). However, it should be noted that, in this review, many more patients were identified with cryptoglandular disease than with CD (1335 patients vs 91 patients).[54]

LIFT is a newer procedure, which has had early success in the treatment of patients with transsphincteric anal fistulae. Use of LIFT in patients with CD has been studied on a limited basis. Similar to other surgical options for patients with anal fistulae related to CD, success rates are lower. A recent prospective study of LIFT in 15 patients with CD revealed a 67% success rate at 12 months. The investigators concluded that patients with anal fistulae associated with CD could be treated with LIFT. No other studies of LIFT in patients with PCD are yet available.[55]

Anal Stenosis and Strictures

Anal stenosis and strictures are a common finding in patients with PCD and can be difficult to treat. Most strictures are thought to be the result of long-standing inflammation.[56] These strictures can be graded as either reversible (inflammatory) or irreversible (fibrostenotic). The location of strictures can be either anal or distal rectal. The location

is important in determining the appropriate treatment. Brochard and colleagues[56] reviewed 102 patients with anorectal strictures in the setting of CD; 76% of strictures were inflammatory and 24% were fibrostenotic. It was observed that 34% patients developed an anal fistula. After a median follow-up of 2.8 years, 59% of the strictures had healed. As would be expected, inflammatory strictures were much more likely to heal. Factors associated with stricture healing were female sex, duration of CD less than 10 years, and presence of an anal fistula; 25% of patients required a stoma. The presence or absence of proctitis was not noted in their study, and it is unknown if this affected healing. The use of biological agents, particularly anti–tumor necrosis factor agents, contributed to the high healing rate of anorectal strictures.[56]

Fecal Diversion for Perianal Crohn's Disease

CD has a bimodal presentation with respect to age of onset. Therefore, it most commonly presents in both younger and older adults.[57] It has been shown that an earlier age at presentation is associated with a more severe phenotype. Also, it follows that more severe disease would require more extensive surgical treatment, including the possibility of a permanent ostomy. Understandably, the prospect of an ileostomy for the younger patient can provoke anxiety.[58]

The prospect of a temporary diverting ileostomy without intestinal resection has been discussed for some time for the patient with severe PCD. A loop ileostomy is the most common form of fecal diversion in this situation, and it can often be done laparoscopically.[59] It has been shown that medical therapy combined with fecal diversion can control the patient'sIBD.[60] One key issue that is important to discuss with the patient preoperatively is whether or not the ostomy is temporary because many of these ostomies may become permanent.

In 2007, Mueller and colleagues[61] reported a series of patients with perianal CD. From 1992 to 1995, 97 patients with CD (whose follow-up data were available) were treated. Of the 97 patients, 51 required temporary fecal diversion (again, most commonly by loop ileostomy). Of the51 patients, 24 ultimately had stoma reversal (47%). The investigators concluded that the risk of permanent fecal diversion was high in patients with complicated perianal disease requiring colorectal resection, but in patients with perianal CD requiring small bowel resection or segmental colonic resection, there was no risk of a permanent stoma.[61]

Mennigen and colleagues[59] identified 33 patients between 2003 and 2012 who underwent what was intended to be a temporary ileostomy for CD; 22 of 29 patients (4 were excluded because of missing data) underwent fecal diversion for perianal CD. Stoma reversal was done in 19 of 25 patients for whom follow-up data was available. However, only 4 of 25 patients had stoma reversal without the need for further surgery. The investigators concluded that although most stomas were indeed temporary, most patients required surgery for the same reason for which they underwent the initial stoma; this was done at a median of 18.5 months after closure of the ileostomy. Of these 19 patients, 7 had a definitive stoma created.[59]

Biological therapy for treatment of IBD has had a significant impact, and treatment of IBD improved with the introduction of infliximab in 1998. Therefore, it would be expected that with improved treatment response rates, the patient with PCD could hope to have a lower risk of a permanent stoma. This fact has been investigated by Hong and colleagues.[62] The investigators identified 21 patients from 1990 to 2007 with perianal CD who underwent fecal diversion. The median age was 34 years. The median follow-up time was 22 months. At 22 months, 4 patients had undergone stoma closure, 11 had had proctocolectomy, and 6 patients still had a stoma. The investigators reviewed the effect of fecal diversion on the course of perianal CD in these

patients. In 4 patients (19%), no effect was seen, and 6 patients had temporary improvement (29%). There was initial improvement with a later plateau in 7 patients (33%) and healing in 4 patients (19%). Of the 21 patients, 11 (52%) received infliximab. In this group, 4 patients underwent proctocolectomy and 2 had intestinal continuity restored. There was no statistically significant difference in stoma reversal between the infliximab and the noninfliximab group.

This view was echoed by Gu and colleagues[63] who reviewed 138 patients with PCD undergoing fecal diversion from 1994 to 2012 at the Cleveland Clinic, Ohio. Only 22% of patients had stoma closure. A total of 45 (33%) patients underwent proctectomy with a permanent stoma and 63 patients (45%) underwent proctectomy with permanent stoma formation after a mean follow-up of 5.7 years. No difference was identified in the outcome based on the type of medical treatment, including treatment with biological agents ($P = .25$).[63] However, when Coscia and colleagues[64] reviewed the course of 233 patients with anorectal CD who were treated between 1995 and 2010, they found that there was a decrease in the risk of permanent stoma from 60.8% in the prebiological therapy era to 19.2% in the biological therapy era. As there seems to be a difference of opinion regarding the impact of biological therapy on the risk of a permanent stoma, debate will most likely continue.

Several different clinical presentations of PCD can make it particularly difficult both to deal with and to treat. The first is the isolated finding of perianal disease without proximal intestinal (either small or large bowel) involvement; this only occurs in less than 5% of patients.[65] The physician who is confronted with unusual perianal disease should recall that CD does not always manifest with intestinal involvement, and this may create difficulty in diagnosis for the treating physician, especially because isolated perianal disease is relatively uncommon. Therefore, it is important for the treating clinician to be aware of isolated perianal CD.

Another presentation that can be difficult to deal with is the presentation of perianal disease in the patient who was previously diagnosed as having UC. While perianal disease is more common in patients with CD, any type of perianal disease, such as anal fissures, anal fistulae, anorectal abscesses, and hemorrhoids can occur in patients with UC. Zabana and colleagues[66] found that 5% of patients with UC had perianal disease. The investigators noted that the diagnosis was changed from UC to CD in one-third of their patients and found a higher requirement for steroids in patients with perianal disease. Therefore, it is uncommon for the patient with UC to have perianal disease. When confronted with the combination of perianal disease and UC, the diagnosis of UC should be questioned and the possibility of CD entertained.

SUMMARY

The management of patients with CD of the colon, rectum, and anus is complex and has changed since the introduction of biological agents. Timing of surgery, the optimal treatment of perianal CD, and the use and avoidance of a stoma are several of the difficult issues in the management of these patients. Segmental colectomy has a role in the management of patients with CD and rectal sparing. Patients with perianal CD should be evaluated for proximal intestinal involvement. Patients with severe perianal CD should have fecal diversion before proctectomy to prevent delayed perineal wound healing.

ACKNOWLEDGMENTS

The author would like to thank Marilyn Teolis for her assistance with researching this article.

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