ABDOMINAL PAIN - INFLAMMATORY BOWEL DISEASE

Epidemiology/Pathophysiology

Inflammatory bowel disease (IBD) is a chronic idiopathic disease, referring to Crohn's disease and ulcerative colitis. It affects approximately 1.4 million Americans and has a peak onset between the ages of 20 to 30 and then a smaller peak at 50-60 years old. IBD is greatest in westernized countries, and in white and Jewish people. In North America, Crohn's disease has an incidence of 319 per 100,000 persons and ulcerative colitis occurs in 249 per 100,000 persons. The increasing incidence of IBD over the 20th century is multifactorial, attributed to environmental factors, increased urbanization, increased awareness, and advancements in diagnostic modalities. There is also a genetic association with IBD, and first-degree relatives have been found to have a 10-fold increased relative risk of developing IBD compared to the general public. Environmental factors that have been associated with IBD include low-fiber diet, food allergies, food additives, and increased sugar. The pathophysiology of IBD is multifactorial, and microbes are identified as a primary contributor. Patients with IBD have increased intestinal epithelial permeability, which allows for a heightened inflammatory response to normal bacterial flora.

Signs and Symptoms

The typical clinical presentation of a patient with IBD is abdominal pain and persistent diarrhea, which is usually bloody, as well as, weight loss and fatigue. Patients with ulcerative colitis often have symptoms of rectal urgency and tenesmus. Other etiologies of colitis must be considered in the differential diagnosis, including infectious and ischemic causes. To help delineate between these diagnoses, patient history must be obtained, pertaining to severity and duration of symptoms, recent travel, antibiotic usage, sexually transmitted disease exposure, ischemic risk factors, family history, and history of pelvic radiation.

Symptoms for IBD patients can be classified as mild, moderate, or severe depending on the amount of abdominal tenderness, and the presence of other systemic symptoms, such as fever, nausea, vomiting, and dehydration. For IBD patients, the disease course consists of varying remissions and flares. The more frequent the flares, the more common the relapses. Patients with Crohn's disease will have perianal disease, such as fissures, fistulas, or abscesses in 20-80% of cases. Patients with ulcerative colitis rarely have perianal disease.

Patients can manifest symptoms related to IBD outside of the gastrointestinal tract. Extraintestinal symptoms can be associated with IBD in 25-40% of patients. The main organ systems affected include eyes, skin, and liver. Common extra-intestinal diseases include ankylosing spondylitis, erythema nodosum, primary sclerosing cholangitis, and iritis/uveitis.

Diagnostic Studies

There is no single test to diagnose IBD. The diagnosis is determined from a combination of symptom history, physical examination, laboratory tests, radiography, and colonoscopy with biopsy.

Physical examination should focus on localizing abdominal tenderness and any signs of peritonitis, such as abdominal rigidity. A rectal examination should be performed as well to evaluate for perianal disease. Patients with Crohn's disease will have perianal disease, such as fissures, fistulas, or abscesses in 20-80% of cases.

Laboratory tests should include a complete blood count, as well as stool culture, including *C. difficile* toxin. Fecal calprotectin has also been identified as a diagnostic tool for IBD, with a sensitivity of 95% and specificity of 91%. Radiographic studies can be performed to aid in diagnosing intestinal complications of IBD. A fluoroscopic exam with a small bowel follow-through can be used to evaluate for small intestinal manifestations, such as stricture.

Endoscopy, either as a colonoscopy or sigmoidoscopy, with biopsy must be performed to definitively diagnose IBD. Grossly, inflammation for Crohn's disease affects all layers and is transmural, and has a cobblestone appearance (Figure 1). In Crohn's disease patients, any portion of the intestinal tract can be affected, although the terminal ileum is the most common, and sparing the rectum. The inflammation is typically patchy and segmental. In contrast, ulcerative colitis extends proximally from the rectum and is continuous. Rectal involvement, also known as proctitis, is a hallmark of the disease. Ulcerative colitis affects only the mucosa and is identified by extensive and diffuse superficial erythema (Figure 2). On histologic biopsy, Crohn's disease forms non-caseating granulomas and ulcerative colitis forms crypt abscesses. Patients with ulcerative colitis are at increased risk of colon cancer, and screening colonoscopies should be performed starting at 8-10 years after initial diagnosis. Surveillance should be continued every 1-2 years.

Non-operative Management

There is no cure for IBD, the treatment is meant to control symptoms and quality of life, as well as decrease the potential complications. The goal of therapy is to attain long-term remission, while providing the least toxic therapy. The management of IBD varies for each type of disease.

Ulcerative Colitis

For ulcerative colitis, the primary management is medical, but 20-30% of patients will ultimately require surgery. The primary treatment of ulcerative colitis consists of managing active disease as well as maintenance therapy. In patients with active disease, the most effective medication is aminosalicylates, such as sulfasalazine and mesalamine. In patients with rectal and left colonic disease, topical formulations, such as suppositories and enemas, are effective in conjunction with oral medications. In patients with severe disease, steroids are the primary

treatment. In patients in whom symptoms are refractory to aminosalicylates and steroids, biologic therapy with infliximab, an anti-TNF medication, has been found to be effective. This is administered as an intravenous infusion over two hours every several weeks. Once the disease is in remission, aminosalicylates are used as the primary therapy for maintenance. Steroids should not be used for long-term maintenance given the side effects. If a patient does not respond well to aminosalicylates, azathioprine can be used, as well as infliximab.

In ulcerative colitis patients with fulminant disease, including symptoms of severe pain, bleeding, and systemic symptoms such as fever and dehydration, inpatient admission is required. The primary therapy in these patients is intravenous steroids. Patients should be followed closely with abdominal exams for the development of toxic megacolon. During these fulminant episodes, any procedure or medications that could exacerbate these symptoms, such as barium enema, colonoscopy, or antidiarrheals, should be avoided.

Crohn's Disease

Crohn's disease typically requires non-operative management. However, within 10 years of diagnosis, 50% of patients will require surgery. Indications for surgery include complications of the disease, including strictures and obstruction. For patients with mild disease, aminosalicylates are used for therapy. Unlike in ulcerative colitis, oral formulations are primarily used, as topical medications are not as effective given the segmental nature of the disease. In patients with moderate to severe disease, steroids are used as the primary treatment. Steroids help induce remission, but do not prevent relapse. Steroids should be tapered off once the symptoms are improved to prevent long-term side effects. If steroids are not effective, biologic therapy with infliximab can be used. Infliximab has an effectiveness of 75-80% in patients. Patients often need to be re-dosed to sustain remission. Aside from infliximab, immunomodulators such as azathioprine and methotrexate can also be used for patients in remission. Immunomodulators affect the immune system in a less specific way than biologic therapy. As in ulcerative colitis, steroids should be avoided for maintenance therapy. Severe and fulminant symptoms often require hospitalization to provide bowel rest, parenteral nutrition, antibiotics, and fluid resuscitation. If symptoms do not improve after 1-2 weeks, surgery is typically required.

Operative Management

Ulcerative Colitis

On average, 25-35% of patients with ulcerative colitis will eventually require surgery, either due to a complication or symptoms refractory to medications. The majority of surgery is elective with a very small amount requiring emergency surgery. Indications for surgery in patients with ulcerative colitis include symptoms intractable to medication, bleeding, toxic megacolon, and cancer.

Symptoms of patients that require emergent surgical intervention include persistent hemorrhage, perforation, and progressive colonic dilation. These patients will have symptoms of bloody diarrhea, weight loss, dehydration, fever, tachycardia, severe anemia, and signs of toxicity. Toxic colitis refers to symptoms including fever, tachycardia, and leukocytosis. Toxic megacolon is distention of the transverse colon to >8cm on abdominal x-ray. Patients with toxic colitis or megacolon require emergent surgical intervention to avoid complications such as perforation. If symptoms do not improve on medication within 1-2 days, surgery will be required. Surgical planning involves removal of the disease and inflamed portion of colon, as an open total or subtotal colectomy with end ileostomy. Total colectomy refers to removal of the entire colon, whereas subtotal refers to a partial removal of the colon. An end ileostomy is when a portion of ileum is brought up through the abdominal wall and sutured to the skin to serve as a conduit of intestinal contents into a pouch. This can be permanent or temporary, and remain in place for 3-6 months after which time the patient can undergo a second operation, an ileostomy reversal, to connect that end of the intestine to the remaining portion of colon, rectum, or anus. Whether the ileostomy is permanent or temporary depends on the extent and severity of patient disease. An ileostomy is performed instead of an anastomosis, to minimize complications such as leak and intra-abdominal infection. Removal of the rectum should not be performed in an emergent setting given the amount of pelvic inflammation.

Elective indications for surgery include colon cancer and medically refractory symptoms, and can usually be performed in an elective setting. These elective operations can be performed minimally invasive, laparoscopically, or robotically, to decrease the hospital length of stay and lead to a faster postoperative recovery. The primary goal of surgery is to remove all the diseased colon and rectum, and restore bowel function, whether in one stage or in subsequent operations. These operations consist of total proctocolectomy, removal of the entire colon and rectum, with end ileostomy, or total proctocolectomy with immediate anastomosis of the ileum to the anus, also known as an ileal pouch-anal anastomosis (IPAA). An IPAA provides patients with a normal mode of defecation. Whether or not patients have an end ileostomy or immediate anastomosis is determined based by the acuity of their symptoms, age, and fecal continence.

Crohn's Disease

The majority of patients with Crohn's disease will require surgery, about 70-90%. Since multiple sites of the small bowel and colon can be affected, patients can present with variable clinical complications. Patients require surgery either for complications or persistent symptoms despite medical therapy. In the emergent setting, patients with Crohn's disease can also develop toxic megacolon, and the management is similar to that described above for ulcerative colitis. Other intestinal complications include abscess, stricture, obstruction, and fistulae. If symptoms do not improve with medical management, surgery is required. Due to the segmental nature of the disease, only the affected area of small bowel or colon should be removed. Additionally, given the chronicity of their disease, only enough bowel should be removed to alleviate current symptoms or complications, as the patient will likely require another operation in the future for another complication. It is important to avoid short-bowel syndrome in these patients, which is when there is not enough functional small intestine to absorb necessary nutrients. On average,

the small intestine is 20 feet, and when it is less than 6.5 feet, patients are more likely to develop short-bowel syndrome. To avoid resection, stricturoplasty can be performed, when the intestine is cut in the direction of the stricture and then re-sutured in the opposite direction to decrease the narrowing. It is also important to optimize a patient's nutrition preoperatively to minimize their risk of postoperative complications.

Perianal disease is present in 20-80% of patients with Crohn's disease. These manifestations include perianal fissures, skin tags, ulcers, abscesses, and fistulae. Control of diarrhea with diet and medications, cleansing the perianal area, and using barrier creams can be helpful to avoid progression of the perianal disease. These conservative attempts are used to manage skin tags and anal fissures. Perianal abscesses and fistulae should not be managed medically and require surgical intervention to avoid perineal sepsis and attempt to close the fistula tracts. These patients will typically require an examination under anesthesia, where the perianal disease can be more closely examined, drained, and probed. If the disease is severe and extensive, and drainage under anesthesia is not adequate to obtain infectious control, patients may require fecal diversion with an ileostomy or colostomy.

Basic Postoperative Care

Postoperative care after colorectal surgery for IBD depends on the acuity of the operation performed and whether an open or minimally invasive approach was performed. In emergent situations, supportive care should be continued, closely monitoring patients for any symptoms or signs of infection. Hospital length of stay is determined by numerous factors, including control of infection, pain control, ability to ambulate independently, diet tolerance, and return of bowel function. The number of days until discharge after surgery is variable. In patients who have an ileostomy, patients should receive education and supplies for stoma site care.

In elective minimally invasive operations, enhanced postoperative recovery pathways have been established to decrease hospital length of stay. Patients typically do not have nasogastric tubes in place after elective colorectal surgery, and are started on a liquid diet within 1-2 days postoperatively. The diet is progressively advanced unless there is evidence of delayed return of bowel function. Urinary catheters may be left in place, but are typically removed after 1-2 days, depending on the extent of dissection performed in the pelvis. IBD patients are typically discharged 3-5 days after elective colorectal surgery for patients.

Questions

- 1. The pathophysiology of inflammatory bowel disease is multifactorial and contributing factors include all of the following except:
 - a. Food additives
 - b. High fiber diet
 - c. Genetic predisposition
 - d. Microbes
- 2. Definitive diagnosis of inflammatory bowel disease is obtained from:
 - a. Fecal calprotectin
 - b. Symptoms including abdominal pain and bloody diarrhea
 - c. Colonoscopy with biopsy
 - d. Rectal examination
- 3. Surgery for inflammatory bowel disease is indicated for patients with:
 - a. At least 5 years of symptoms related to the disease
 - b. Colon polyps
 - c. Symptoms refractory to medical management
 - d. Infectious colitis

Answers

- 1. The pathophysiology of inflammatory bowel disease is multifactorial and contributing factors include all of the following except:
 - a. Food additives
 - b. High fiber diet
 - c. Genetic predisposition
 - d. Microbes

The pathophysiology of inflammatory bowel disease is multifactorial. Contributing factors include a genetic association with first-degree relatives having a 10-fold increased relative risk of developing the disease compared to the general public, and environmental factors including low-fiber diet, food allergies, food additives, increased sugar, and microbes.

- 2. Definitive diagnosis of inflammatory bowel disease is obtained from:
 - a. Fecal calprotectin
 - b. Symptoms including abdominal pain and bloody diarrhea
 - c. Colonoscopy with biopsy
 - d. Rectal examination

The diagnosis of inflammatory bowel disease is determined from a combination of symptom history, physical examination, laboratory tests, radiography, and colonoscopy with biopsy. Endoscopy, either as a colonoscopy or sigmoidoscopy, with biopsy must be performed to definitively diagnose inflammatory bowel disease.

- 3. Surgery for inflammatory bowel disease is indicated for patients with:
 - a. At least 5 years of symptoms related to the disease
 - b. Colon polyps
 - c. Symptoms refractory to medical management
 - d. Infectious colitis

Surgery for inflammatory bowel disease is indicated in the emergent setting, with persistent hemorrhage, perforation, and progressive colonic dilation. Electively, surgery is performed for concern of colon cancer, symptoms refractory to medical therapy, and complications such as obstruction, recurrent abscesses, and fistulae.

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Figures

Figure 1. Gross cobblestone appearance of Crohn's disease on colonoscopy

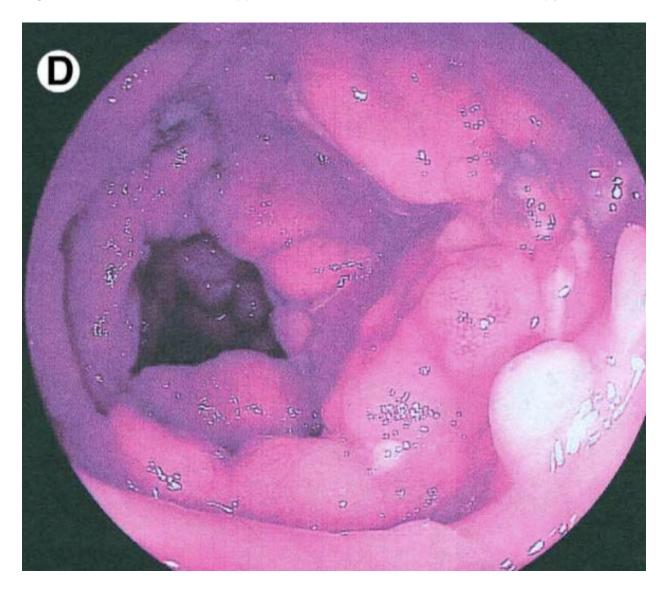
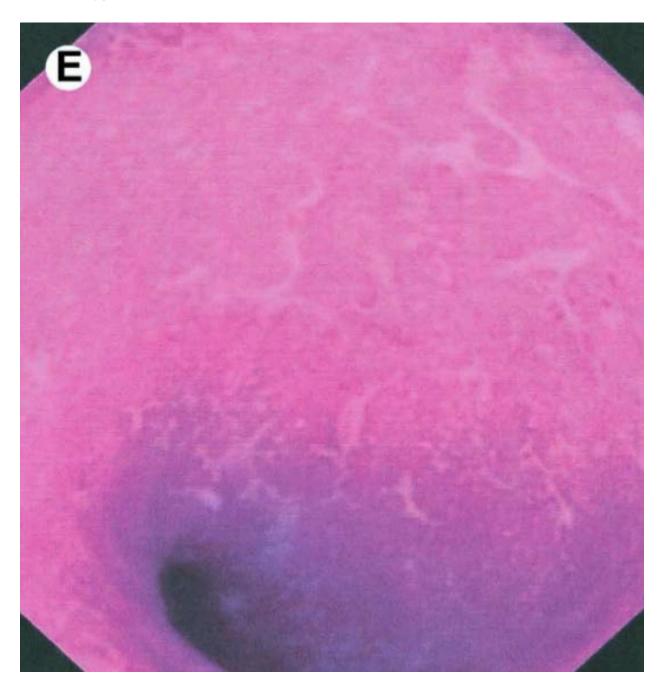


Figure 2. Superficial erosions and erythema, appearance of ulcerative colitis on colonoscopy



Author

Rana Higgins, MD, FACS, FASMBS
Froedtert Hospital and Medical College of Wisconsin, Milwaukee, WI