Pediatric Inflammatory Bowel Disease In The Emergency Department: Managing Flares And Long-Term Complications

Abstract

Inflammatory bowel disease includes both Crohn disease and ulcerative colitis. Pediatric-onset inflammatory bowel disease differs from adult inflammatory bowel disease in disease type, location, progression, and sex preponderance, and 20% to 30% of inflammatory bowel disease is diagnosed in childhood. Children are more likely than adults to present with extraintestinal manifestations of inflammatory bowel disease (with aphthous ulcers, joint involvement, and growth delay being the most common). Inflammatory bowel disease flares typically require treatment with intravenous steroids and inpatient admission. Acute emergencies include toxic megacolon, intestinal obstruction, and perforation. The use of steroids may obscure diagnosis of an underlying abdominal emergency by masking signs and symptoms. The emergency clinician must be cognizant of such complications and diagnostic challenges when evaluating inflammatory bowel disease.
### Case Presentations

A previously healthy 12-year-old boy is brought to the ED by his mother for evaluation of fatigue and skin rash. She has noticed “red bumps” on his legs for the past few weeks, but she reports no other associated symptoms. The patient says he has been feeling tired lately and can’t seem to keep up with his friends on the playground. He states that he sometimes has abdominal cramping after eating but denies diarrhea. He does not recall being bitten by insects on his legs and says those red bumps “hurt just a little.” On physical examination, you note a thin and mildly pale-appearing boy, who is small for his age. He has normal affect and behavior. Vital signs are: oral temperature, 36.3°C; heart rate, 89 beats/min; respiratory rate, 18 breaths/min; blood pressure, 103/65 mm Hg; and oxygen saturation, 99% on room air. Several well-circumscribed, minimally tender, nonweeping and nonblanching erythematous nodules are noted on the bilateral anterior tibia. There is no surrounding fluctuance, crepitus, or bullae. The only significant finding on review of his past medical history is his growth chart. You are concerned that the nodules on his legs may be indicative of systemic disease, and you consider your diagnostic options...

You are then called to evaluate a 17-year-old adolescent girl who is brought in by ambulance for abdominal pain and fever. She has a history of Crohn disease and is currently taking sulfasalazine and prednisone. She reports diffuse abdominal cramping for 2 days, fever and chills, vomiting, and bloody diarrhea. She has been unable to tolerate any oral intake since 3:00 AM that day. She denies any recent antibiotic use, history of travel, or medication changes, although she has not been consistently compliant with her medications. Vital signs are: temperature, 39.1°C; heart rate, 132 beats/min; respiratory rate, 23 breaths/min; blood pressure, 94/57 mm Hg; and oxygen saturation, 95% on room air. She appears pale and diaphoretic, and her mucous membranes are dry. Physical examination is notable for diffuse abdominal tenderness with guarding, tympany on percussion, and decreased bowel sounds. No rebound tenderness is elicited. Capillary refill is 3 seconds. The remainder of the examination is within normal limits. You ask your nursing staff for 2 large-bore intravenous lines for fluid resuscitation. You are concerned about an acute flare and you consider appropriate treatment options...

### Critical Appraisal Of The Literature

A literature search was performed using PubMed, Ovid MEDLINE®, Google Scholar, and the Cochrane Database of Systematic Reviews with the search terms pediatric inflammatory bowel disease, epidemiology, emergency, treatment, complications, and malignancy. Over 200 articles met the selection criteria. Of those, 109 articles with full texts were reviewed, and 87 are cited in this review. Clinical cohort and systematic review studies were also analyzed. Few prospective trials have been conducted for treatment in the pediatric population. Further research is needed in the management of the child with IBD.

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**Inflammation**

Inflammatory bowel disease (IBD) includes both Crohn disease and ulcerative colitis. Approximately 2 million people worldwide are afflicted with IBD, and 20% to 30% of all patients with IBD are diagnosed during childhood.\(^1,2\)

Childhood incidence of ulcerative colitis is estimated at 0.5-4.3/100,000 and Crohn disease at 0.2-8.5/100,000.\(^3\) The peak incidence of initial presentation for IBD occurs between the ages of 15 and 25 years, and approximately 20% of patients with ulcerative colitis and 25% to 30% of patients with Crohn disease present before the age of 20 years.\(^4\) Crohn disease and ulcerative colitis occur equally in the first 8 years of life, but Crohn disease is more common in older children.\(^5\) The incidence of ulcerative colitis has remained relatively stable, whereas the incidence of Crohn disease has increased.\(^5-9\) Utilization of colonoscopy in developed countries may have led to greater differentiation of Crohn disease from ulcerative colitis and relatively more diagnoses of Crohn disease. The number of emergency department (ED) visits per year is unknown; however, the public health burden of disease is significant in patients with IBD, due to utilization of outpatient resources, ED visits, and inpatient care.\(^10\)

Certain clinical presentations may be similar to adults, but pediatric-onset IBD differs in disease type, location, progression, and sex preponderance. While there is no difference in sex predominance in adult IBD, studies have shown that Crohn disease is more prevalent in pediatric male populations.\(^11\) Importantly, a subset of children with IBD may present only with extraintestinal manifestations (particularly joint involvement, apthous ulcers, poor weight gain, growth failure, and delayed puberty). Of all extraintestinal manifestations, joint involvement is the most common in children with IBD.\(^12\)

Both medical and surgical interventions have the goal of inducing and maintaining remission in IBD. However, these treatments are not without side effects, the most significant of which are immunosuppression, infusion reaction, and postsurgical complications.

Potential complications of IBD include intestinal obstruction and perforation, sepsis, and toxic megacolon. Long-term disease burden includes an increased risk of developing malignancies and recurrent thromboembolic events.
Etiology And Pathophysiology

The exact pathophysiology of IBD is not well understood. There is likely a combination of environmental, genetic, and immunologic factors. An environmental trigger may incite a dysregulation of the immune response to gut flora in a genetically susceptible host.\textsuperscript{13-15} Patients with IBD have less complex profiles of commensal bacteria and higher numbers of mucosa-associated bacteria as compared to healthy individuals.\textsuperscript{13} Increased intestinal permeability and food-borne bacterial infections trigger some IBD cases.

Though the cause of IBD is thought to be multifactorial, there are many known risk factors.

Genetic Susceptibility

A family history of IBD carries an 8- to 10-fold increased risk, and IBD occurrence in first-degree relatives may be as high as 40%.\textsuperscript{3,16} In one report, children of parents with IBD (either Crohn disease or ulcerative colitis) had up to a 33% chance of developing IBD before 30 years of age.\textsuperscript{17}

Ethnicity

IBD has a higher incidence in Caucasians and those of Jewish descent. The disease course is particularly severe in Jewish persons with Familial Mediterranean Fever.\textsuperscript{18}

Infection

\textit{Clostridium difficile} infection can be either an inciting or exacerbating factor.\textsuperscript{19} An increased risk of IBD has been shown in persons after acute gastroenteritis with \textit{Salmonella} or \textit{Campylobacter} infections.\textsuperscript{20-21} The risk is highest during the first year after the gastroenteritis episode, but it remains high for up to 15 years compared to matched controls.\textsuperscript{21}

Environment

Smoking, exclusive formula-feeding, increased stress, and a diet rich in animal protein, total fat, and polyunsaturated fatty acids\textsuperscript{22} are associated with increased risk of IBD.\textsuperscript{3} The incidence of IBD has historically been higher in industrialized, developed countries. A recent increase in incidence globally suggests that environmental factors play a significant role, and such epidemiological evolution is linked to the modern Westernized lifestyle.\textsuperscript{23}

Differential Diagnosis

The differential diagnosis largely depends on the clinical presentation of each individual patient (See Table 1); infectious, immune-related, and vascular etiologies, as well as acute abdomen mimics, must be considered.

Prehospital Care

Emergency stabilization should be initiated promptly by emergency medical services, particularly if the patient exhibits signs and symptoms of dehydration or hypoperfusion (such as hypotension, delayed capillary refill, changes in mental status, cool and mottled skin, and respiratory distress). Large-bore intravenous access should be obtained, and fluid resuscitation should be utilized to correct vital sign abnormalities. It is also important for emergency medical services personnel to gather the patient’s prior treatment information and medications at home, if available.

<table>
<thead>
<tr>
<th>Presentation Symptom</th>
<th>Differential Diagnosis</th>
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<tbody>
<tr>
<td>Bloody diarrhea</td>
<td>• Infectious colitis (\textit{Shigella}, \textit{Salmonella}, \textit{Yersinia spp.}, \textit{Campylobacter}, \textit{C difficile}, enterohemorrhagic \textit{Escherichia coli})  \n</td>
</tr>
<tr>
<td>Watery diarrhea</td>
<td>• Parasitic infection (cryptosporidiosis, giardiasis)  \n</td>
</tr>
<tr>
<td>Rectal bleeding</td>
<td>• Anal fissure/polyp  \n</td>
</tr>
<tr>
<td>Perirectal disease</td>
<td>• Fissure  \n</td>
</tr>
<tr>
<td>Abdominal pain (particu-\textsuperscript{larly in the right lower quadrant)}</td>
<td>• Appendicitis, +/- periappendiceal abscess  \n</td>
</tr>
<tr>
<td>Growth delay</td>
<td>• Endocrinopathy  \n</td>
</tr>
</tbody>
</table>

Emergency Department Evaluation

In general, a thorough head-to-toe physical examination should be performed, including skin and mucosal surface.

Clinical Presentation

Pediatric patients with IBD can present to the ED with signs and symptoms of colitis. Often, they have a subacute illness with abdominal pain and bloody diarrhea. Fever, anemia, fatigue, nausea, vomiting, and weight loss are also common presenting symptoms in IBD. Symptoms may be acute, persistent (≥ 4 weeks), or recurrent (≥ 2 episodes in 6 months). Extraintestinal manifestations occur in 6% to 10% of children upon presentation, with a total incidence of 25% to 35%. Inflammatory bowel disease flares can be precipitated by bacterial, parasitic, or viral superinfection. Among such superinfection, C difficile and cytomegalovirus are the most common and most challenging to manage.

Indeterminate Colitis

Younger children (aged < 8 years) with IBD are more likely to have isolated colonic disease. As the diagnosis of either ulcerative colitis or Crohn disease can initially be unclear, children are frequently labeled as having indeterminate colitis. There is no widely accepted definition of indeterminate colitis; thus, this includes a heterogeneous subset of patients. Indeterminate colitis has been reported in 3% to 30% of pediatric IBD.

Ulcerative Colitis

Ulcerative colitis is limited to the colon and involves the superficial mucosa. The mucosa is friable, and bleeding may occur spontaneously. Lesions are continuous and start from the rectum. Although rare, with severe disease, ulcerations may develop and penetrate the muscularis layer. At presentation, 44% to 49% of children with ulcerative colitis have rectosigmoid disease, 36% to 41% have left-sided disease, and 14% to 27% have pancolitis. Childhood ulcerative colitis tends to be more severe as compared to adults. Approximately 80% to 90% of children either present with or progress to pancolitis, in contrast to only 40% to 50% seen in adults. There is a shorter time from diagnosis to first surgery in children with ulcerative colitis as compared to adults. One study demonstrated that, at 10 years after diagnosis, 40% of childhood onset ulcerative colitis had required a colectomy versus 20% of adult-onset ulcerative colitis. In the majority of patients, the clinical course involves disease flares alternating with periods of remission. A minority of patients have continuous disease activity. Overall, disease activity tends to decrease over time.

Crohn Disease

Crohn disease is marked by transmural bowel wall inflammation that occurs throughout the gastrointestinal tract, but it can occur anywhere from the mouth to the anus. Normal mucosa surrounds diseased segments (often described as “skip lesions”). In both children and adults, Crohn disease most commonly affects the terminal ileum and the proximal colon. Younger children predominately have colonic disease, though the upper gastrointestinal tract is involved in up to two-thirds of children with Crohn disease.

The most common presenting symptoms of Crohn disease include abdominal pain, diarrhea, rectal bleeding, and weight loss. Crohn disease often has a more insidious onset, and patients present with vague complaints of malaise and mild abdominal discomfort. Extraintestinal manifestations (including arthralgia and growth and sexual development delays) can dominate the clinical picture and may even be present in children prior to gastrointestinal symptoms. Thus, diagnostic delays are common.

The clinical course of pediatric Crohn disease tends to be more severe compared to adults. The course is unpredictable with recurrent exacerbations of symptoms and, more rarely, continuous active disease. Ulcerations may become deep and develop complications, such as abscesses, fistulae, strictures, and obstruction from adhesions. Small bowel disease results in obstruction and colonic involvement, often presenting with bleeding (most commonly in the form of hematochezia). There is no direct correlation between symptoms and progression of anatomic damage. A small segment of ileitis can cause severe abdominal pain and fatigue, while strictures and fistulas may be asymptomatic for years.

Extraintestinal Symptoms

Extraintestinal manifestations associated with IBD are seen in approximately 6% to 10% of patients at presentation, and they can be seen in up to 30% of patients within a few years after being diagnosed. Occasionally, patients may present with extraintestinal symptoms before any gastrointestinal symptoms develop. These symptoms include oral superficial ulcerations (aphthous stomatitis), episcleritis/iritis, arthralgia/arthritis, skin disease (erythema nodosum and pyoderma gangrenosum), and nonspecific mild elevations of serum aminotransferases (a common finding, but often transient and may relate to medications or disease activity). Growth failure and sexual development delay are common in the pediatric population. (See Table 2, page 5.)
Diagnostic Studies

There is no consensus regarding the diagnostic criteria for IBD. The diagnosis is often established by examining a patient’s presenting symptoms and clinical features along with laboratory blood testing and imaging studies. Endoscopy with histopathologic examination of biopsied tissue samples is the gold standard of diagnosis; and it can also provide further differentiation between Crohn disease and ulcerative colitis.

Laboratory Testing

Although normal laboratory test results do not exclude the diagnosis of IBD, recommended initial tests include complete blood count (CBC), complete metabolic panel, and inflammatory markers (such as C-reactive protein [CRP] and erythrocyte sediment rate [ESR]). Lipase and liver function tests should be included as well. Type and screen should be considered, as blood transfusion may be necessary. Urinalysis should also be sent to rule out a genitourinary etiology. Common abnormalities include anemia (microscopic or normocytic), thrombocytosis, increased ESR and CRP (particularly in Crohn disease) and mild elevation of aspartate aminotransferase (AST) and alanine aminotransferase (ALT). Low albumin is commonly present because of malnutrition in children with IBD. Laboratory findings are more likely to be abnormal to a greater extent with severe disease.

In patients presenting with diarrhea (particularly bloody diarrhea), stool culture should be sent for enteric pathogens, such as Salmonella, Shigella, Yersinia, Campylobacter, C difficile, Giardia, E coli, Entamoeba, and tuberculosis. The incidence of C difficile is greater in children when compared to adults and frequently correlates with severity of disease in young children. In severe colitis, blood and stool cultures (including evaluation for C difficile and other enteric pathogens) should be obtained to evaluate for concurrent infection. Studies have shown that 5% to 25% of patients with ulcerative colitis flares have underlying C difficile infection and have a worse outcome.

The most widely used diagnostic panel in the United States is the IBD Serology 7 panel® (Prometheus laboratories, San Diego, CA). Anti-Saccharomyces cerevisiae antibody (ASCA) and perinuclear antineutrophil cytoplasmic antibody (p-ANCA) are disease-specific for Crohn disease and ulcerative colitis, respectively. Many new serologic markers are currently under investigation for both the diagnosis of IBD and for differentiating between Crohn disease and ulcerative colitis. One such marker, fecal calprotectin, has been found to be significantly elevated in all patients with IBD. Serologic markers can also be applied to stratify IBD patients with respect to disease progression.

Diagnostic Imaging

Patients with IBD in an acute flare typically present with abdominal pain and/or distention, along with bloody diarrhea and fever. An acute abdominal x-ray series may be used as an initial test in the ED to evaluate for serious complications of IBD. It should include an upright chest x-ray to examine for free air, an upright abdominal film to examine for air-fluid levels, and a supine abdominal film to view the width of the bowel loops that are most visible. The abdominal series provides a rapid screening tool to evaluate for the presence of bowel obstruction or perforation as well as toxic megacolon.

On the acute abdominal series x-ray, signs of bowel obstruction (such as air-fluid levels, a paucity of distal bowel gas, or free air suggestive of perforation) may be present. Toxic megacolon appears as colonic dilatation with an abnormal mucosal contour, typically most pronounced in the transverse colon. Acute dilatation (to > 5 to 6 cm) of the transverse colon with loss of haustral folds in a severe exacerbation of colitis is diagnostic in older children and adults, with 90% sensitivity and 90% specificity reported in one study. In children aged < 10 years, transverse colonic diameter > 4 cm is suggestive of toxic megacolon.

Table 2. Extraintestinal Symptoms Of Inflammatory Bowel Disease

<table>
<thead>
<tr>
<th>Extraintestinal Manifestations</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arthritis</td>
<td>Occurs in 7% to 25% of patients, Non erosive and asymmetric, Large joints are involved (hips, knees, wrist)</td>
</tr>
<tr>
<td>Abdominal</td>
<td>Cholelithiasis, pancreatitis, primary sclerosing cholangitis, May result in bile duct scarring and cirrhosis</td>
</tr>
<tr>
<td>Erythema nodosum, pyoderma gangrenosum</td>
<td>Occurs in 5% to 10% of patients and resolves with inflammatory bowel disease treatment</td>
</tr>
<tr>
<td>Nephrolithiasis</td>
<td>Due to calcium malabsorption and increased oxalate uptake, Occurs in 5% of patients</td>
</tr>
<tr>
<td>Iritis, uveitis</td>
<td>Photophobia, red eye, vision changes</td>
</tr>
<tr>
<td>Stomatitis, aphthous ulcers</td>
<td>May improve with inflammatory bowel disease treatment, Occurs in &lt; 1% of patients, More common in adults</td>
</tr>
<tr>
<td>Growth delay</td>
<td>Short stature may result, Delayed sexual development, May be the only symptom prior to diagnosis of inflammatory bowel disease</td>
</tr>
<tr>
<td>Osteoporosis</td>
<td>Calcium malabsorption, malnutrition, corticosteroid use</td>
</tr>
</tbody>
</table>
megacolon.\textsuperscript{40} However, clinical appearance of illness is much more telling than imaging.

In addition to x-ray, computed tomography (CT) may be necessary to evaluate for other complications (including fistula, abscess, or perforation). Such complications should be suspected in toxic-appearing patients, patients with severe and/or escalating abdominal pain, and patients with signs and symptoms of peritonitis. Typically, contrast agents are used for better evaluation of the bowel wall and lumen as well as for detecting the presence of masses or obstructions. Oral contrast can be given even in the setting of suspected perforation, but water-soluble contrast agents (such as Gastrografin\textsuperscript{\textregistered}) should be used, as these agents do not provoke an inflammatory response if leaked into the peritoneal cavity.

Magnetic resonance imaging (MRI) is used more commonly in pediatric IBD as it avoids ionizing radiation exposure, although this technique is less well-established.\textsuperscript{41-44} Ultrasound is not currently used for IBD screening, but it may play a larger role in the future as the use of ultrasound for gastrointestinal conditions becomes more established.\textsuperscript{45} Ultrasound may be used in an acute exacerbation and can detect intestinal hypervascularity, lymphadenopathy, and thickened bowel loops.\textsuperscript{46} Mesentery can be evaluated, and fluid collections and abscesses may be visualized. Advanced ultrasound techniques are emerging using oral, rectal, and even intravenous contrast to improve scan quality. Ultrasound offers the benefits of not exposing the child to radiation and being relatively inexpensive and accessible, although it is highly operator-dependent.

When deciding which imaging modality should be pursued, consider the patient’s presentation, appearance, and clinical examination findings. Such decisions should also be made in conjunction with pediatric gastrointestinal and surgical consultants, and the center-specific availability of the imaging modalities should be considered.

**Outpatient Imaging**

Potential treatment options and workup can include outpatient imaging. Emergency clinicians should be knowledgeable of these procedures when discussing treatment options with patients.

**Endoscopy**

Colonoscopy is performed in patients with suspected IBD, and biopsies are taken for histopathologic analysis. Many gastroenterologists often concomitantly perform upper endoscopy to differentiate between Crohn disease and ulcerative colitis. While each has specific features seen on endoscopy, there is frequent overlap between Crohn disease and ulcerative colitis in children. Diffuse colonic involvement is common in both pediatric ulcerative colitis and Crohn disease, and both may have gastric ulcers.\textsuperscript{47}

**Crohn Disease**

Ileal inflammation is present in most Crohn disease cases. Non-necrotizing granulomas confirm the diagnosis of Crohn disease, but they are only present in approximately 50% to 60% of patients.\textsuperscript{48,49} Cobblestoning, described as mucosal ulcerations interlaced with normal segments of bowel (skip lesions), bowel wall edema, bowel lumen narrowing or obstruction, and enteric fistulas may be seen. Relative rectal sparing is frequently seen.

**Ulcerative Colitis**

Diffuse and continuous inflammation that starts at or near the rectum and extends proximally into the colon is indicative of ulcerative colitis. Early in pediatric ulcerative colitis, atypical features may occur with patchy disease and relative rectal sparing. Non-specific inflammation in the upper gastrointestinal tract may occur in up to 75% of children.\textsuperscript{29} Pseudo-polyps, which are areas of regenerating mucosa from prior areas of ulceration, are often present.

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**Figure 1. Abdominal X-ray Showing Toxic Megacolon**

Upper Gastrointestinal Contrast Study
In addition to endoscopy, imaging of the upper gastrointestinal tract (esophagus, stomach, small intestine) using contrast is recommended to further differentiate between Crohn disease and ulcerative colitis. Traditionally, an upper gastrointestinal contrast study with small bowel follow-through has been the standard for evaluation. With advancing technology, alternative modalities are available and are being increasingly utilized. In a stable patient, such work-up is typically deferred to outpatient, with referral to a pediatric gastroenterologist.

Video-Capsule Endoscopy
Video-capsule endoscopy is used in adults to evaluate Crohn disease severity. It is approved by the FDA for patients aged > 10 years but has been used in children as small as 11.5 kg with an acceptable complication rate. The most observed complication of video-capsule endoscopy is retained capsule with subsequent bowel obstruction and perforation. The benefit is the avoidance of radiation, sedation, and general anesthesia.

Treatment

Management Overview
The overall goal in IBD management for the treating clinicians as well as for emergency clinicians is to induce remission and minimize flares. The selection of 1 or more therapeutic agents is guided by the severity and extent of gastrointestinal and extragastrointestinal manifestations. Maintenance medications are used, with additional medication interventions as needed, for acute flares. The effectiveness of various therapies is difficult to assess because there are few adequately powered, high-quality pediatric studies available. The current published data include studies with methodological shortcomings and are frequently cohort studies. Exclusive enteral nutrition in which the patient only receives elemental formula (via mouth, nasogastric tube, or gastrostomy tube) as the sole source of nutrition has been shown to effectively induce remission in children with IBD. This therapy is more common in Canada and Western Europe and is used less commonly in the United States. Adherence is difficult, and exclusive enteral nutrition may be required for 6 to 8 weeks to induce remission.

Management Of Inflammatory Bowel Disease

Acute Flares
Severe acute flares are characterized by > 6 bloody stools per day, abdominal pain/distension, fever, tachycardia, anemia, and inflammatory changes on laboratory values. The inflammatory response from IBD alone may cause a low-grade fever. The challenge for the emergency clinician is differentiating between an acute disease flare and a more serious complication (such as infectious colitis, toxic megacolon, bowel obstruction, or perforation). Additionally, the use of corticosteroid or immunomodulatory medications may mask a patient’s symptoms.

Acute flares require intravenous fluids and typically intravenous steroids. High-dose steroids (methylprednisolone at 1 mg/kg/dose every 12 hours to a maximum of 30 mg every 12 hours) are most commonly used. Occasionally, oral steroids may be used if patients are not toxic-appearing, although such a decision should be made in conjunction with pediatric gastrointestinal consultants.

Bowel Perforation And Toxic Megacolon
Bowel perforation is more common in ulcerative colitis than Crohn disease, especially when occurring with toxic megacolon. The incidence of bowel perforation in IBD is estimated to be 1% to 3%, although a more recent study indicates that the incidence can be as high as 16%. Escalating abdominal pain, evidence of peritonitis, and a patient’s ill appearance should alert the emergency clinician to the possibility of toxic megacolon and/or perforation. Toxic megacolon occurs in approximately 5% of adults with IBD, but it is less common in children. It is a surgical emergency and is associated with an increased risk of perforation, sepsis, electrolyte abnormalities, and hemorrhage.

Bowel Obstruction
Bowel obstruction is a common complication in IBD. Patients classically present with nausea, vomiting, obstipation, and abdominal pain with distention. Air-fluid levels are seen on abdominal series x-ray. When bowel obstruction is suspected, patients should be placed on bowel rest with intravenous hydration. It is important to note that, while nasogastric tube decompression has traditionally been utilized as part of conservative treatment, one study found that its routine use in patients without active emesis is associated with respiratory failure and pneumonia.

Other Management Options
Suspected infectious colitis, toxic megacolon, or perforations are indications for antibiotics. Severe disease or complications may require surgical intervention. Adult data have shown that 67% of patients with a severe ulcerative colitis flare respond to intravenous steroids, and 29% require colectomies. Pain control remains a challenge in IBD. Opioid use is associated with toxic megacolon, particularly in active colitis, so the use of other pain medications should be considered. Some of the literature recommends low-dose morphine (0.05 mg/kg intravenously). Benzodiazepine use for tenesmus and ketamine (0.1-0.5 mg/kg) for pain control have also been reported.
Clinical Pathway For Management Of Pediatric Inflammatory Bowel Disease

Patient presents with:
- Abdominal pain/distention
- +/- bloody diarrhea
- +/- extraintestinal symptoms

History of IBD?

Yes

Toxic-appearing?

No

- Consider differential diagnosis (See Table 1, page 4)
- Pain control
- PMD/GI follow-up
  (Class II)

Yes

- Blood culture
- Stool culture
- IV fluids
- IV steroids
- Consider IV antibiotics
- GI consultation
- Admit
  (Class II)

If acute abdomen:
- Imaging
- Surgical consultation
  (Class II)

No

Toxic-appearing?

Yes

- Consider differential diagnosis (See Table 1, page 4)
- Pain control
- Laboratory workup
- PMD follow-up
- Consider GI referral
  (Class II)

No

- Consider differential diagnosis (See Table 1, page 4)
- Pain control
- Laboratory workup
- IV fluids
- IV steroids
- Consider IV antibiotics
- GI consultation
- Admit
  (Class II)

- Blood culture
- Stool culture
- IV fluids
- IV steroids
- Consider IV antibiotics
- GI consultation
- Admit
  (Class II)

Abbreviations: GI, gastrointestinal; IBD, inflammatory bowel disease; IV, intravenous; PMD, preferred medical doctor.

Class Of Evidence Definitions

Each action in the clinical pathway section of Pediatric Emergency Medicine Practice receives a score based on the following definitions.

Class I
- Always acceptable, safe
- Definitely useful
- Proven in both efficacy and effectiveness

Level of Evidence:
- One or more large prospective studies are present (with rare exceptions)
- High-quality meta-analyses
- Study results consistently positive and compelling

Class II
- Safe, acceptable
- Probably useful

Level of Evidence:
- Generally higher levels of evidence
- Non-randomized or retrospective studies: historic, cohort, or case control studies
- Less robust randomized controlled trials
- Results consistently positive

Class III
- May be acceptable
- Possibly useful
- Considered optional or alternative treatments

Level of Evidence:
- Generally lower or intermediate levels of evidence
- Case series, animal studies, consensus panels
- Occasionally positive results

Indeterminate
- Continuing area of research
- No recommendations until further research

Level of Evidence:
- Evidence not available
- Higher studies in progress
- Results inconsistent, contradictory
- Results not compelling

This clinical pathway is intended to supplement, rather than substitute for, professional judgment and may be changed depending upon a patient's individual needs. Failure to comply with this pathway does not represent a breach of the standard of care.

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Medications
Many classes of medications can be used to treat flares of IBD in the ED. Emergency clinicians should also be aware of the medications used for long-term treatment of IBD, and should obtain this information when taking the patient’s history. A summary of typical IBD medications and side effects is provided in Table 3.

Antibiotics
Antibiotics are not indicated in the treatment of acute colitis unless underlying infectious colitis is suspected. Distinguishing between an acute flare and infectious colitis may be difficult, and emergency clinicians must rely on a detailed history and physical examination. While the type of antibiotics needed is dependent on the underlying infectious organism, the initiation of antibiotics should be correlated with a patient’s clinical examination. Typically, ciprofloxacin (Cipro®) and metronidazole (Flagyl®) combination therapy is used for 10 to 14 days as first-line therapy. Emergency clinicians should be mindful that, while systemic fluoroquinolones are not frequently used as first-line therapy in pediatric patients younger than adolescence, they are considered a reasonable alternative for situations where no safe and effective substitute is available.

Topical Therapy
Oral lesions may be treated with topical prednisolone syrup (5 mg/5 mL) or dexamethasone (Dexamethasone Intensol®) (0.5 mg/5 mL), either applied directly to lesions or by the swish-and-spit method twice daily. If the lesions are localized to the lips, triamcinolone 0.1% may be used 2 to 4 times per day. If disease is limited to the rectum, topical therapy is used. Rectal hydrocortisone enemas or foam or triamcinolone 0.1% may be used 2 to 4 times per day. If the lesions are localized to the lips, methasone Intensol® (0.5 mg/5 mL), either applied directly to lesions or by the swish-and-spit method twice daily. If the lesions are localized to the lips, triamcinolone 0.1% may be used 2 to 4 times per day. If disease is limited to the rectum, topical therapy is used. Rectal hydrocortisone enemas or foam or triamcinolone 0.1% may be used 2 to 4 times per day. If the lesions are localized to the lips, methasone Intensol® (0.5 mg/5 mL), either applied directly to lesions or by the swish-and-spit method twice daily. If the lesions are localized to the lips, triamcinolone 0.1% may be used 2 to 4 times per day. If disease is limited to the rectum, topical therapy is used. Rectal hydrocortisone enemas or foam or triamcinolone 0.1% may be used 2 to 4 times per day.

Corticosteroids
In both pediatric Crohn disease and ulcerative colitis, corticosteroids are used for induction of remission and flares, but they are not for maintenance of remission. Steroids are used for 2 to 4 weeks and then tapered off to diminish side effects, risk of developing adrenal insufficiency, and growth disturbances. Oral corticosteroids are used in mild disease (prednisone 1 to 1.5 mg/kg/day, maximum 40 to 60 mg/day). Oral corticosteroids are escalated to intravenous steroids (methylprednisolone 1 mg/kg per dose every 12 hours, max 60 mg/day) if the patient is unresponsive to oral corticosteroids after 1 to 2 weeks of treatment. Intravenous steroids are used initially for severe disease.

Aminosalicylates
5-ASA is used for both induction and maintenance of remission in pediatric IBD, though there are currently no randomized controlled trials published on children. Examples of this class of medication include sulfasalazine (Azulfidine®), mesalamine (Apriso®, Pentasa®), and olsalazine (Dipentum®). 5-ASA agents inhibit the synthesis of proinflammatory prostaglandins and leukotrienes in the colon. Mild exacerbation of watery diarrhea is common during the first few weeks of use of 5-ASA preparations, but initial worsening of colitis symptoms (cramps, diarrhea, and rectal bleeding) indicates an adverse reaction, which most commonly occurs with mesalamine. These patients are typically termed “allergic” to 5-ASA preparations. It is important to note that 5-ASA agents are ineffective during an acute exacerbation and should be discontinued during a flare, as the use of 5-ASA agents may paradoxically worsen colitis symptoms. As sulfasalazine inhibits folic acid transport on the cellular level, folic acid supplementation is recommended in all patients taking sulfasalazine to avoid megaloblastic anemia.

<p>| Table 3. Medications And Side Effects Of Treatment For Inflammatory Bowel Disease |
|-----------------------------------------------|-------------------|-----------------|</p>
<table>
<thead>
<tr>
<th>Medication Class</th>
<th>Specific Agents</th>
<th>Side Effects</th>
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<tr>
<td>Corticosteroids</td>
<td>Prednisone</td>
<td>Accelerated atherosclerosis</td>
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<td></td>
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<td>Adrenal insufficiency (hypothalamic-pituitary-adrenal axis suppression)</td>
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<td></td>
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<td>Gastritis, pancreatitis</td>
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<td>Growth delay</td>
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<td>Hypertension</td>
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<td>Immunosuppression</td>
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<td>Mood disorder/psychosis</td>
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<td>Osteoporosis</td>
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<td></td>
<td>Skin thinning/purpura</td>
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<tr>
<td>Aminosalicylates</td>
<td>5-ASA</td>
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<td></td>
<td>Mesalazine</td>
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<td></td>
<td>Olsalazine</td>
<td>Rash</td>
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<td></td>
<td>Sulfasalazine</td>
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<td>Hemolytic anemia</td>
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<td>Leukopenia</td>
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<td>Thrombocytopenia</td>
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<td></td>
<td></td>
<td>Pancreatitis</td>
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<tr>
<td>Immunomodulator</td>
<td>AZA, 6-MP</td>
<td>Leukopenia</td>
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<td></td>
<td></td>
<td>Liver toxicity</td>
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<tr>
<td>Methotrexate</td>
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<td>Gastrointestinal intolerance (nausea, vomiting, loose stool)</td>
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<td>Hepatotoxicity</td>
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<td>Steromatosis</td>
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Abbreviations: 5-ASA, 5-aminosalicylic acid; AZA, azathioprene; 6-MP, 6-mercaptopurine.
Immunomodulators

Though varying in mechanism of actions, 6-mercaptopurine (6-MP) (Purinethol®, Purixan®) and azathioprine (AZA) (Azasan®, Imuran®) show efficacy in inducing remission in ulcerative colitis and maintaining remission in pediatric Crohn disease. Both 6-MP and AZA have been shown to induce remission in 70% to 80% of steroid-dependent Crohn disease, but may take 2 to 4 months to work. Methotrexate (Trexal®) has quicker onset of action, but there are only small trials of its use in children. It is important to note that AZA and 6-MP have narrow therapeutic windows, and their levels must be routinely monitored to avoid bone marrow and liver toxicity. Thiopurine methyltransferase enzyme activity is a major factor in determining AZA and 6-MP metabolism. This enzyme’s activity level should be checked on an outpatient basis in all patients in whom AZA or 6-MP is being considered.

Biologic/Anti-Tumor Necrosis Factor Agents

Infliximab (Remicade®) is the most commonly used biologic agent and acts directly against the cytokine tumor necrosis factor-alpha (TNF-alpha). It is used both to induce and maintain remission in IBD resistant to steroids and immunomodulators. Adalimumab (Humira®) has a similar mechanism of action, but it also functions as another option for patients who have lost response to or have developed adverse reactions to infliximab. Success rates are high, but a patient may require repeated doses and treatment for a long period of time. Biologic agents are typically dosed every few weeks to months. Long-term side effects of immunosuppression place patients at risk for sepsis, opportunistic infections, and lymphoma. An infusion reaction occurs in 15% to 25% of patients receiving biologic agents. Initial symptoms are similar to anaphylaxis (including laryngospasm, shortness of breath, acute chest discomfort, mucosal irritability, and hemodynamic instability). Delayed symptoms occur 2 to 14 days after an infusion and include fever, arthritis/arthritis, myalgias, myalgias, malaise, and urticarial rash. Premedication does not prevent initial infusion reaction but may decrease subsequent reactions. Reactions are typically managed with epinephrine, antihistamines, corticosteroids, and decreased infusion rate. Many of the medications used to treat IBD can lead to immunosuppression. Therefore, screening for infectious disease (such as tuberculosis and human immunodeficiency virus) should be considered prior to starting such therapy.

Surgery

It has been estimated that as high as 70% to 90% of patients with Crohn disease, and 25% to 30% of patients with ulcerative colitis, will require surgery at some point in the disease course. Surgical intervention is reserved for patients with IBD that has not responded to medical therapy or for certain complications (such as an abscess, fistula or stricture). Surgery is also indicated in acute emergencies, such as uncontrolled hemorrhage, bowel perforation, and toxic megacolon. In patients with ulcerative colitis, if disease is limited to the colon, a colectomy essentially “cures” the disease. However, studies have shown that disease relapses and bowel re-resections are common during the first decade after primary surgery. Endoscopic balloon dilation with steroid injection treatment is used as an alternative to surgery. Success rates are higher in patients with simple strictures.

Due to the high incidence of surgical complications for patients with IBD, knowledge of these complications and the recommended management is imperative for the emergency clinician. Overall, the incidence of surgical complications in children is between 13% and 55%. Early postsurgical complications include wound infections, dehiscence, pouch ischemia, anastomotic leakage, and perforation. Late complications include pouchitis and fistula formation. Intestinal obstruction may occur initially or years after the initial surgery. Ileal pouchitis is the major complication following ileo-anal pull-through and restorative proctocolectomy, and occurs in 10% to 44% of patients. The majority of patients have a single initial attack after surgery. Symptoms include acute onset of diarrhea (possibly > 30 episodes/day), urgency, incontinence, rectal bleeding, abdominal cramping, and malaise. Endoscopic examination of the pouch demonstrates mucosal hemorrhage, ulcers, and contact bleeding in the ileal mucosa. Extrapileal manifestations of colitis often relapse with pouchitis. Treatment includes antibiotics (typically metronidazole) and corticosteroid enemas. Most cases resolve within 72 hours of treatment. Chronic pouchitis is less common. The etiology of pouchitis is unknown, but theories include ischemia, bacterial overgrowth, immunologic abnormality, and recurrent disease in cases where colonic metaplasia develops.

Long-Term Complications Of Inflammatory Bowel Disease

As with any other chronic disease, both physical and psychosocial burdens imposed on patients are substantial, especially in the pediatric population. Long-term complications of IBD include the risk of developing malignancy, thromboembolic risks, adverse medication effects, reduced growth velocity, and psychosocial implications. In IBD overall, there is a small increase in mortality of 0.5% over age-matched controls. Mortality is primarily due to infections (particularly in patients receiving immu-
nosuppressive agents), followed by malignancy and uncontrolled disease. Difference in clinical outcome between sexes has not been observed.

**Malignancy**
The risk of colorectal cancer in patients with IBD has decreased in the past 30 years, but those with pancolitis, early onset of IBD (< 25 years of age), and the male sex are at increased risk. The risk of dying from colon cancer is estimated to be 8% after 10 years from the diagnosis of IBD. Cancer has been reported in children aged < 18 years. Surveillance for cancer by colonoscopy is recommended. Epithelial dysplasia generally precedes carcinoma; therefore, regular screening for dysplasia should be done. Aside from colorectal cancer, patients with IBD are also at an increased risk of developing malignancy of biliary tract, skin, and hematologic origins.

**Thromboembolic Risks**
IBD is a well-established risk factor for recurrent venous thromboembolism. The pathophysiology behind this is not completely understood. It is likely multifactorial and frequently related to acquired risk factors (such as surgery, immobilization, dehydration, and indwelling central venous lines).

**Medication Complications**
Immunosuppression from various agents increases the risk of disseminated and opportunistic infections. Steroid use can often mask signs and symptoms of acute abdomen (such as toxic megacolon and perforation), leading to increased morbidity and mortality. Use of azathioprine and mercaptopurine increases risk for nonmelanoma skin cancers. Use of immunomodulators increases risk of lymphoma. A rare fatal form of lymphoma, hepatosplenic T-cell lymphoma, is associated with the use of 6-MP and AZA with or prior to the use of biologic agents.

**Growth**
Pediatric IBD has unique manifestations with respect to the growth and development of a child. Consequences of IBD include malnutrition, failure to thrive, delayed puberty, and short stature. Proinflammatory cytokines exert deleterious effects on growth either systemically or at the level of the growth plate. Defective bone mass accrual and low bone density are also seen, and they are strongly associated with growth failure.

**Psychosocial Effects**
The psychosocial effects of a developing child with IBD can be significant and complex. Fear of strangers, separation anxiety, fear of loss of love and approval, fear of loss of control of bodily functions, and fear of pain and humiliation are common in children.

Studies have shown that up to 25% of adolescents with IBD are depressed. Social support, education, and skills to cope with stress are important in caring for a child with IBD. Online resources for teens include [www.ibdu.org](http://www.ibdu.org) and [www.ccfa.org](http://www.ccfa.org). These are resources an emergency clinician can give patients and caregivers.

**Other Long-Term Complications**
Premature subclinical atherosclerosis may occur in pediatric IBD. There are early reports that the microvascular endothelial dysfunction, which occurs in IBD, is similar to that observed in lupus and rheumatoid arthritis. Other complications of IBD include nephrolithiasis (due to malabsorption of calcium and increased absorption of oxalate), cholelithiasis (due to bile acid malabsorption), and osteopenia (which may be caused by vitamin D deficiency and/or by treatment with glucocorticoids). Granulomatous inflammation of other areas (including the lymph nodes, gonads, and lungs) may also be seen, particularly in patients with Crohn disease.

**Disposition And Summary**
Inflammatory bowel disease should be considered in a child or adolescent presenting with abdominal pain and/or distention, diarrhea, perianal disease, or any extraintestinal manifestations involving the skin, joints, bones, or eyes. Inquire about the child’s growth and puberty as well as the parental growth patterns and age at onset of puberty for comparison when IBD is suspected. Pediatric patients with suspected IBD should be referred to a pediatric gastroenterologist for further workup, if consultation is not expected. Laboratory testing includes CBC, complete metabolic panel, and inflammatory markers (CRP and ESR). While anemia, hypoalbuminemia, and elevated CRP and ESR are frequently observed in either the initial presentation and/or in an acute flare, normal laboratory results do not rule out IBD.

An underlying acute surgical abdomen must be considered in a patient with known or suspected IBD presenting to the ED with acute symptoms (such as abdominal pain, distention, bloody diarrhea, fever, or vomiting). It is often difficult to determine whether the patient is having an acute disease flare or is developing complications (such as intestinal obstruction, perforation, or toxic megacolon). Active investigation should occur for underlying infection. Acute abdominal series x-ray should be obtained as an initial test to look for air-fluid levels, free air, and/or the presence of toxic megacolon. Advanced imaging, such as ultrasound, CT, or MRI (if available) should be considered when suspicion is high for intestinal obstruction, perforation, or abscess. Intravenous fluids for underlying dehydra-
tion and high-dose steroids for disease exacerbation should be given. In addition to the aforementioned laboratory testing, blood and stool cultures should be sent to look for the presence of enteric pathogens. Intravenous antibiotics are indicated in suspected infectious colitis, toxic megacolon, or perforation. Surgical intervention may be necessary, and early surgical consultation is recommended.

Corticosteroids are the most effective method for inducing remission. They may be used topically for relatively localized disease or orally. In an acute flare, intravenous steroids are required. Other medications include aminosalicylates, immunomodulators, and biologic agents. Antibiotics are typically indicated if there is underlying infectious colitis, as some pediatric gastroenterologists do use ciprofloxacin and/or metronidazole on a temporary basis as adjunctive treatment in acute flares.

Patients with known IBD may have many "classically adult" complications and may present to the ED with these complaints. Nephrolithiasis, thromboembolism, diseases associated with accelerated atherosclerosis, and malignancies are some of the associated afflictions. Additionally, patients are often immunocompromised due to medication regimens. This results in increased infection risk as well as the potential to mask an underlying surgical abdomen. The complications and diagnostic challenges of pediatric IBD that have been reviewed here will ensure that emergency clinicians can provide the highest level of care to patients.

Risk Management Pitfalls For Inflammatory Bowel Disease
(Continued on page 13)

1. “You can’t diagnose IBD when patients don’t have any GI symptoms.”
   Although IBD typically manifests with abdominal pain/distention and diarrhea that is often bloody, it can also present with only extraintestinal manifestations. These are particularly prominent in the pediatric population (especially arthralgias, delayed puberty, and delayed growth).

2. “I am worried about an intra-abdominal abscess, but I decided not to order CT of the abdomen in this pediatric patient because the radiation risk is too high.”
   There are various imaging modalities that could aid in the diagnosis of IBD or in the detection of IBD-related complications. Such imaging modalities include x-ray, CT, MRI, ultrasound, and endoscopy. The choice of imaging modality largely depends on the patient’s clinical presentation, consideration of risks versus benefits, and availability of the chosen modality. If the patient appears toxic and the clinical examination is concerning for possible bowel perforation and/or toxic megacolon, then abdominal CT may provide the necessary information in the shortest period of time. Computed tomography with oral contrast can evaluate the bowel wall and lumen, and it can identify perforation, obstruction, and abscesses. If using oral contrast and there is a concern for perforation, use a water soluble contrast medium.

3. “If all laboratory testing results are normal (including inflammatory markers), then this patient does not have IBD.”
   Normal laboratory test results do not exclude the diagnosis of IBD, but laboratory testing should be done when IBD is suspected. Common abnormalities seen in laboratory testing include anemia (usually normocytic or microcytic), thrombocytosis, elevated ESR and CRP, mild elevation of AST and ALT, and hypoalbuminemia. In addition, studies have shown that red blood cell distribution width is markedly increased in active IBD and may be useful in monitoring disease progression.

4. “I saw a pediatric patient with painful oral ulcers. She has a history of IBD and is taking multiple immunosuppressants. She appeared well otherwise, so I sent her home with ibuprofen only.”
   In addition to pain medication, such oral lesions may be treated with topical prednisolone syrup (5 mg/5 mL) or dexamethasone (0.5 mg/5 mL), either applied directly to the lesions or by the swish-and-spit method twice daily. If the lesions are localized to the lips, triamcinolone 0.1% may be used 2 to 4 times per day.

5. “Patients with IBD always need antibiotics when they present with abdominal pain.”
   Antibiotics are indicated when there is suspicion of infectious colitis, toxic megacolon, or intestinal perforations. Blood and stool cultures should be sent prior to initiating antibiotics.
Case Conclusions

After reviewing the 12-year-old boy’s growth chart, you noticed that he had crossed below the growth curves for both weight and stature. You remembered that growth delay can sometimes be the only presenting symptom of inflammatory bowel disease, which caused you to suspect that those red bumps were actually erythema nodosum. You decided to send off the following laboratory tests: complete metabolic panel, CBC, CRP, and ESR. The results were consistent with your suspected diagnosis, as the patient had microcytic anemia, hypoalbuminemia, and an elevated CRP. Since this patient did not have abdominal pain or diarrhea and was nontoxic-appearing, you decided not to pursue any imaging or to start antibiotics. You discussed the suspected diagnosis with the patient’s mother and made a referral to a pediatric GI specialist for further workup and outpatient endoscopy.

For the 17-year-old, you decided to place the following orders: complete metabolic panel, CBC, CRP, ESR, amylase, urinalysis, pregnancy test, and KUB x-ray. You prescribed antiemetics and antipyretics, but you withheld pain control due to the possible linkage between opioids and toxic megacolon in patients with IBD. The patient stopped vomiting following antiemetic administration but continued to complain of diffuse abdominal pain. You decided to trial 0.3 mg/kg of ketamine, which seemed to alleviate her discomfort. Repeat vital signs were: blood pressure, 106/63 mm Hg; heart rate, 110 beats/min; respiratory rate, 18 breaths/min; temperature, 38.2°C orally; and oxygen saturation, 98% on room air. When the KUB came back, it showed a segment of abnormally dilated distal transverse colon as well as absence of haustra and thickened colonic walls. You did not see any air-fluid levels or free air. Con-

Risk Management Pitfalls For Inflammatory Bowel Disease

(Continued from page 12)

6. “I can’t tell if the patient is having an acute flare or having complications secondary to Crohn disease, so I was unsure of the best way to treat the patient.”

It is often difficult to distinguish between acute flares or IBD complications solely based on physical examination and laboratory testing, as findings can be very similar in both circumstances. Imaging can aid in differentiating these 2 etiologies; however, it may not be needed because management is similar for both conditions. Patients often require intravenous steroids, intravenous fluid hydration, and broad-spectrum intravenous antibiotics (if underlying infection is suspected).

7. “This parent is asking for a flu shot for her child, but the boy is taking immunomodulators for IBD.”

Patients taking immunomodulators have suppressed immune systems and are more prone to developing infections. Sepsis is one of the leading causes of mortality in patients with IBD. Thus, it is important to vaccinate patients. Immunization with inactivated vaccines (particularly influenza, pneumococcus, and meningococcus) should be maintained and updated. Depending on the level of immunosuppression, live attenuated vaccines (Measles, Mumps, and Rubella; Varicella; intranasal influenza) may be contraindicated. Decisions regarding whether specific vaccinations are appropriate for patients on immunomodulatory medications for IBD should be made by the prescribing clinician.

8. “X-ray findings of toxic megacolon are the same in adults as in children.”

Findings of toxic megacolon in both adults and children include colonic dilatation with an abnormal mucosal contour, which is typically most pronounced in the transverse colon. Acute dilatation of the transverse colon to > 5 to 6 cm with loss of haustral folds during a severe exacerbation of colitis is diagnostic of toxic megacolon in older children and adults. In children aged < 10 years, transverse colonic diameter > 4 cm is suggestive of toxic megacolon.

9. “This patient is having a flare of IBD, but I wasn’t sure if all outpatient medications should be continued.”

During a flare, patients typically require high dose intravenous steroids. The decision to continue other classes of outpatient medication (such as biologic agents or immunomodulators) should be made in conjunction with gastrointestinal specialists. It is important to note that 5-ASA agents (sulfasalazine, mesalamine, olsalazine) are ineffective during an acute exacerbation and should be discontinued to prevent worsening of colitis symptoms.

10. “This patient looks like he is having an acute flare, but I was concerned about further immunosuppression, so no steroid was given.”

While many patients with IBD are relatively immunocompromised due to immunomodulator therapy, intravenous steroids are indicated as one of the essential treatments during an acute flare. High-dose steroids (methylprednisolone 1 mg/kg/dose every 12 hours to a maximum of 30 mg every 12 hours) are most commonly used.
cerned about toxic megacolon, you immediately placed a call to your surgical and GI colleagues for inpatient admission and possible surgical intervention. You also wrote the following orders: broad-spectrum intravenous antibiotics, blood and stool cultures, high-dose intravenous steroids, NPO, and normal intravenous saline.

References

Evidence-based medicine requires a critical appraisal of the literature based upon study methodology and number of subjects. Not all references are equally robust. The findings of a large, prospective, randomized, and blinded trial should carry more weight than a case report.

To help the reader judge the strength of each reference, pertinent information about the study will be included in bold type following the reference, where available. In addition, the most informative references cited in this paper, as determined by the authors, will be noted by an asterisk (*) next to the number of the reference.


Time-And Cost-Effective Strategies

- **Antibiotic use:** Antibiotics are only indicated when there is suspicion of infectious colitis, toxic megacolon, or intestinal perforation. In a patient with a history of IBD presenting with abdominal pain with or without bloody diarrhea, but who is otherwise well-appearing and has a benign abdominal examination, antibiotics may not be necessary. It is worthwhile to educate both patients and their parents on side effects and microbial resistance associated with overuse of antibiotics.

- **Imaging:** Imaging can provide added information when emergency clinicians are concerned about intestinal obstruction, perforation, megacolon, and/or abscess. There are many imaging modalities, each with varying sensitivity depending on underlying pathology. The imaging benefit versus risk must be weighed and the modality selected should be the one that provides the greatest amount of information for the clinical question.


22. Inflammatory bowel disease in children and adolescents:


4. Which of the following statements is TRUE when comparing Crohn disease to ulcerative colitis in pediatric patients?
   a. Both Crohn disease and ulcerative colitis can present as colitis, but only ulcerative colitis can progress to pancolitis.
   b. Crohn disease involves the superficial mucosa and is limited to the colon.
   c. Skip lesions are much more common in Crohn disease than ulcerative colitis.
   d. Ulcerative colitis is much more common in older children.

5. Which of the following statements regarding pediatric Crohn disease in FALSE?
   a. Abscess, fistula, and stricture development are common.
   b. Crohn disease is marked by continuous lesions involving the mucosal surface from mouth to anus.
   c. Extraintestinal manifestations are common and can be the only presenting symptoms.
   d. Younger children predominately have colonic disease with less involvement of the upper gastrointestinal tract.

6. You are called to evaluate a 16-year-old adolescent girl who was brought to the ED by her parents for abdominal pain, vomiting, and diarrhea. She has a history of Crohn disease and is currently taking azathioprine. She has been having bloody diarrhea for the past 2 days, along with subjective fever, chills, and abdominal pain. Vital signs are: temperature, 38.8°C; heart rate, 134 beats/min; respiratory rate, 22 breaths/min; blood pressure, 98/66 mm Hg; and oxygen saturation, 94% on room air. Physical examination of the abdomen reveals diffuse tenderness with rigidity on palpation, rebounding, and guarding. Which of the following imaging modalities would be the most appropriate for initial screening?
   a. Acute abdominal series x-ray
   b. CT of the abdomen/pelvis with contrast
   c. MRI with contrast
   d. Ultrasound

1. Which of the following risk factors has NOT been associated with the development of pediatric inflammatory bowel disease?
   a. Coexisting diagnosis of atopy
   b. Family history of IBD
   c. Infection with *Campylobacter*
   d. Smoking

2. Paramedics bring in a 16-year-old adolescent boy from a clinic for shortness of breath and chest pain. He has a history of ulcerative colitis and was at the clinic receiving his scheduled IV medication. On arrival to the ED, his vital signs are: temperature, 37.7°C; heart rate, 151 beats/min; respiratory rate, 34 breaths/min; blood pressure, 78/56 mm Hg; and oxygen saturation, 88% on 15-L nonrebreather mask. Which of the following is least appropriate in the management of this patient’s condition?
   a. Epinephrine
   b. Normal saline bolus
   c. Packed red blood cell transfusion
   d. Steroids

3. When comparing pediatric- and adult-onset inflammatory bowel disease, which of the following statements is TRUE?
   a. Both Crohn disease and ulcerative colitis tend to have a more severe clinical course in adults than in children.
   b. Crohn disease most commonly affects the terminal ileum in children.
   c. Pancolitis is much more common in the pediatric population.
   d. Time from diagnosis to first surgery is shorter in adults with ulcerative colitis.
7. Paramedics bring in a 17-year-old adolescent boy with a history of Crohn disease for fever and abdominal pain. According to the patient’s mother, he has been having abdominal pain, > 10 episodes of bloody diarrhea per day, and fever for the past 2 days. He has not had any recent illness or travels. He is currently taking sulfasalazine. Vital signs are: temperature, 39.5°C; heart rate, 127 beats/min; respiratory rate, 26 breaths/min; blood pressure, 95/68 mm Hg; and oxygen saturation, 96% on room air. Physical examination reveals diffuse abdominal tenderness without rebound, decreased bowel sounds, and frank blood on stool guaiac examination. Which of the following is least appropriate in the management of this patient’s condition at this time?
   a. Broad-spectrum antibiotics
   b. Continue current medication
   c. Corticosteroids
   d. Low-dose ketamine

8. For the patient mentioned in question 7, which of the following statements regarding this patient's condition is TRUE?
   a. Intravenous corticosteroids may worsen the clinical condition with further immune suppression.
   b. The initial phase of this condition includes fever, myalgia, and rash.
   c. Patients may progress into a delayed phase of this condition with laryngospasm, acute chest pain, and hemodynamic instability.
   d. Premedication does not prevent the occurrence of the medication reaction.

9. Which of the following medications is least effective in maintaining remission of pediatric IBD?
   a. Azathioprine
   b. Infliximab
   c. Olsalazine
   d. Prednisone

10. Regarding long-term complications of pediatric inflammatory bowel disease, which of the following statement is FALSE?
   a. Delayed puberty and osteopenia are common.
   b. Earlier age at onset of disease is associated with increased risk of developing colorectal cancer.
   c. Primary sclerosing cholangitis occurs most commonly in patients with ulcerative colitis.
   d. Recurrent venous thromboembolism has not been reported despite increased risk of malignancy development.
In Volume 1 of the Pediatric Emergency Medicine Audio Series, Dr. Andrew Sloas reviews 4 critical pediatric topics and provides evidence-based treatment recommendations.

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Physician CME Information

Date of Original Release: July 1, 2014. Date of most recent review: June 15, 2014.

Accreditation: EB Medicine is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians. This activity has been planned and implemented in accordance with the Essential Areas and Policies of the ACCME.

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Needs Assessment: The need for this educational activity was determined by a survey of medical staff, including the editorial board of this publication; review of morbidity and mortality data from the CDC, AHA, NCHS, and ACEP; and evaluation of prior activities for emergency physicians.

Target Audience: This enduring material is designed for emergency medicine physicians, physician assistants, nurse practitioners, and residents.

Goals: Upon completion of this activity, you should be able to: (1) demonstrate medical decision-making based on the strongest clinical evidence; (2) cost-effectively diagnose and treat the most critical ED presentations; and (3) describe the most common medicolegal pitfalls for each topic covered.

CME Objectives: Upon completion of this article, you should be able to: (1) distinguish the differences between pediatric- and adult-onset inflammatory bowel disease, as well as the differences between Crohn disease and ulcerative colitis; (2) describe the medical and surgical treatments for inflammatory bowel disease; and (3) identify and manage complications of inflammatory bowel disease.

Discussion of Investigational Information: As part of the newsletter, faculty may be presenting investigational information about pharmaceutical products that is outside Food and Drug Administration approved labeling. Information presented as part of this activity is intended solely as continuing medical education and is not intended to promote off-label use of any pharmaceutical product.

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