A 7-year-old Boy with a History of Intermittent Bloody Diarrhea
Robert Listernick, MD

A 7-year-old boy was admitted for evaluation of a 4-month history of intermittent bloody diarrhea. Initially, he had formed stools with streaks of blood on the outside. He was seen by his doctor, who diagnosed constipation. Over the next 4 months, he started having progressively looser stools that became bloody and watery. He had been having at least 10 watery stools each day for the last month with generalized abdominal pain. There was no history of prolonged fevers, recurrent emesis, rash or arthritis.

Past history is remarkable for two febrile seizures as an infant. Family history was remarkable for a paternal uncle with peptic ulcer disease. There was no significant travel history.

On exam, he was an alert, interactive, pale boy. Pulse was 102, temperature 98.6°F, respiratory rate 16, blood pressure 107/72. Weight was in the 80th percentile and height in the 95th percentile. HEENT exam was unremarkable. Lungs were clear. Abdomen was soft, without tenderness, masses or organomegaly. He had Tanner 1 genitalia. The testes were descended. There was no peripheral edema or digital clubbing. Neurologic exam was normal.

Laboratory evaluation: Hemoglobin 8.6 g/dL, MCV 69; white blood cell count 8,000/mm³ with 63% neutrophils, 34% lymphocytes; platelet count 550,000/mm³. Serum chemistries were normal save for albumin 3 g/dL. Erythrocyte sedimentation rate was 64 mm/hour.

Robert Listernick, MD, moderator: What should be the pediatrician’s approach to the evaluation of hematochezia?

Barry Wershil, MD, pediatric gastroenterologist: Certainly, if the hematochezia is acute with streaks of blood on the stool itself, you’d want to make sure that the child is not constipated. However, before any interventions are performed, if the child has diarrhea, a stool culture should be obtained to rule out the possibility of a bacterial gastroenteritis with the more common organisms.

Robert Tanz, MD, general academic pediatrician: Certainly this testing is well within the domain of the general pediatrician, long before subspecialty referral is necessary. I would also look at his growth chart for evidence of poor weight gain or linear growth as evidence of chronicity. If he has acute bloody diarrhea, and he’s developing hemolytic uremic syndrome, for instance, referral to a gastroenterologist would be inappropriate.

Dr. Listernick: If the general pediatrician suspects inflammatory bowel disease (IBD), is there a role for radiology testing before referral to a subspecialist?

Dr. Wershil: Given that endoscopy and colonoscopy are routine procedures, there’s little reason to perform a barium enema or a CT if IBD is the primary concern.

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Dr. Listernick: Can you comment on the utility of the various IBD serology panels?

Dr. Wershil: Various serologic panels test for antibodies against components of yeast, certain bacteria, and neutrophils, a combination of which may yield some predictive ability to identify the presence of ulcerative colitis (UC) or Crohn’s disease (CD). These antibodies titers are placed into a logistical mathematical model to predict the likelihood of IBD and the type of IBD. There are specific instances where they can be useful. However, I cannot recommend that this test be ordered by a general pediatrician. For example, 50% of children who have IBD have negative serologic testing.

It’s possible that in the future, subsets of patients will be identified by this technology who may be more prone to aggressive disease or respond better to a particular class of drugs, but we are not there yet.

Dr. Listernick: The patient underwent colonoscopy and biopsy. Can we see the initial biopsies?

Elaine Cham, MD, pediatric pathologist: Looking at biopsy specimens of the colon, we see a striking degree of inflammation with increased cellularity in the lamina propria and multiple crypt abscesses with collections—that is, neutrophils within the crypts.

In addition, there is architectural distortion with irregularity in the size and distribution of the crypts. The terminal ileum biopsies appeared normal. These findings are virtually continuous from the ascending colon to the rectum and are highly suggestive of UC.

Dr. Wershil: Unfortunately, despite all our advances, no single test distinguishes between CD and UC; the ultimate diagnosis is made by the physician utilizing all data at hand—clinical testing, radiography, laboratory testing, and pathology. Even the presence of granulomas, highly suggestive of CD, may be seen in some infections, as well as in the colitis associated with chronic granulomatous disease.

In this child, the sum of the data strongly suggest UC, particularly with the terminal ileum being normal. To absolutely assure ourselves that he doesn’t have any small intestinal disease outside of the terminal ileum, which would mitigate against the diagnosis of UC, we should perform either a barium study with small bowel follow-through or capsule endoscopy.

The problem with capsule endoscopy is that one can’t get biopsies. Remember that pathology is very important in distinguishing between UC and CD. Many younger gastroenterologists tend to overinterpret the capsule endoscopy findings without getting histologic confirmation.

Dr. Listernick: Initial treatment?

Dr. Wershil: With this degree of involvement, I would start intravenous corticosteroids. Between 80% and 90% of patients will undergo dramatic improvement, usually in the first week, and can be discharged on oral corticosteroids. As they are weaned from the steroids, as many
as 50% relapse, depending upon the age and extent of disease.

**Dr. Listernick:** Would earlier diagnosis make a difference?

**Dr. Wershil:** That’s a hard question to study. However, the historical interval reported between onset of symptoms and initial IBD therapy has been 14 months; more recently, we’ve been able to decrease that interval to 6 to 8 months.

**Dr. Listernick:** I’m going to briefly summarize the next part of the history. He experienced marked improvement initially with the development of normal bowel movements and increased appetite and energy. He gained 1.5 kg over the next 6 weeks. However, several months later, he was found to have hemoglobin of 7 g/dL, despite being relatively asymptomatic. Repeat endoscopy revealed diffuse erythema and exudates with inflamed mucosa and many pseudopolyps throughout the entire colon.

He was started on infliximab. Biopsy samples were negative for cytomegalovirus (CMV), Epstein-Barr virus (EBV), and adenovirus by polymerase chain reaction.

Two months later, he began having severe daily headaches and was seen by an optometrist, who noted the presence of bilateral papilledema. His mother immediately took him to an emergency room, where he had a normal CT scan of the head. Lumbar puncture showed an opening pressure of 36 cm of water, but normal CSF cell count, glucose, and protein.

**Dr. Listernick:** Why did they look for viruses in this highly inflamed colonic mucosa?

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Ellen Chadwick, MD, pediatric infectious disease physician: CMV is well-known to cause colitis in immunocompromised individuals. I wouldn’t have tested for EBV or adenovirus. The best way to diagnose CMV colitis is to look for the presence of giant cells on biopsy. A positive CMV PCR might simply indicate the presence of CMV in the biopsy specimen, which wasn’t an active participant in the disease process.

**Stanford T. Shulman, MD, pediatric infectious disease physician:** I should point out that amebic colitis may be misdiagnosed as IBD if the pathologist doesn’t carefully exclude the presence of amoebae in biopsy specimens or stool.

**Dr. Listernick:** So why does this child have increased intracranial pressure?

**Leon Epstein, MD, pediatric neurologist:** Although there is common “wisdom” that poor vision causes headaches, I’ve yet to see such a child. That should not be the pediatrician’s kneejerk reaction.

In this case, he had clear evidence of increased intracranial pressure without hydrocephalus, so-called pseudotumor cerebri, or idiopathic intracranial hypertension. Characteristic headaches of pseudotumor cerebri tend to wake patients up at night or are worse in the morning due to hypoventilation and minor rises in retained carbon dioxide during sleep, which leads to cerebral vasodilatation.

Treatment with, and withdrawal from, corticosteroids are known to cause increased intracranial pressure; the mechanism is not clear, although it probably is the result of decreased CSF reabsorption by the arachnoid granulations.

**Dr. Listernick:** What about infliximab?

**Dr. Wershil:** Infliximab is an antitumor necrosis factor antibody that markedly decreases inflammation. It can be useful as maintenance therapy of IBD in steroid-resistant patients. The response rate for patients with all forms of IBD is approximately 80%, although less than 60% have a sustained remission when infliximab is discontinued. These individuals often end up undergoing colectomy.

**Dr. Listernick:** Does colectomy ever occur early in the course?
**Dr. Wershil:** Remarkably, a recent study documented that 9% of children with UC undergo colectomy at time of initial presentation. Furthermore, an additional 20% of patients require colectomy in the first year following diagnosis. The risk of adenocarcinoma of the colon in patients with UC starts to rise dramatically after 20 years of disease activity; cancer is not really a pediatric problem.

**Kathy Barsness, MD, pediatric surgeon:** There are several different indications for colectomy with restorative ileoanal J-pouch. As mentioned, cancer prophylaxis is a big indication in the patient who has had UC for more than 20 years, particularly once you start seeing dysplasia on biopsies. Fulminant, refractory colitis is another indication.

The goals of surgery are to eliminate the disease by removing segments of the intestine with the exception of 1 or 2 cm at the anal verge and to restore normal bowel functional anatomy and allow bowel movements through the anus.

**Dr. Listernick:** Can you describe the operation or series of operations that are performed?

**Dr. Barsness:** We rarely perform a single stage operation in patients with IBD because of their inflammation and poor wound healing secondary to the immunosuppressive drugs that are used. The first stage is to perform a colectomy, leaving a short segment of sigmoid and rectum down at the peritoneal reflection, and create a J-pouch. After this has healed, we can take down the ileostomy and restore bowel function through the anus.

**Dr. Listernick:** J-pouch?

**Dr. Barsness:** We take the end of the ileum and partially fold it back up on itself for approximately 12 cm, creating the short end of the J. The long end of the J is the rest of the small intestines. A pouch is created and sewn to the remaining rectal muscles, which creates a reservoir for stool; the other end of the J becomes an ileostomy. After about 2 months, allowing the bowel to heal, we reconnect the bowel to the anus. Most surgeons would do this laparoscopically.

**Dr. Listernick:** What is their functional outcome?

**Dr. Barsness:** Adolescents tend to do better than older adults. Adolescents may have four to six somewhat looser bowel movements each day. We hope the stools have a thick, pudding-like consistency, rather than being more watery. We often use loperamide to help achieve that consistency. Over time, the remaining bowel may increase its water-reabsorbing capacity.

**Dr. Listernick:** Is there any morbidity from the colectomy itself?

**Dr. Barsness:** Fertility rates drop by 20% to 40%. We do our best to manage women medically during the child-bearing years before performing colectomy for this reason.

**Dr. Wershil:** Even though surgery in UC may be looked on as a failure of medical therapy by some individuals, surgery may significantly increase the quality of life and is an important therapeutic option in our armamentarium. I’ve seen many children who look at us with profound relief and gratitude following colectomy; their pain is gone for the first time in years.

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### Key Learning Points

1. In the initial diagnostic approach to bloody diarrhea, stool culture should be performed.
2. If inflammatory bowel disease is suspected, there’s little reason to perform barium enema or computerized tomography (CT) scans of the abdomen. The initial diagnostic procedure should be colonoscopy and, in some situations, esophagogastroduodenoscopy.
3. Both treatment with and withdrawal from corticosteroids are known to cause increased intracranial pressure (pseudotumor cerebri).
4. Decreased fertility in women is a significant long-term complication of colectomy.