A 12-year-old Girl with Bloody Diarrhea

By Dorota Walkiewicz-Jedrzejczak, MD

Educational Objectives
After reading this article and answering the review questions, the reader will be able to:

1. Recognize the signs and symptoms suggestive of Inflammatory Bowel Disease (IBD)
2. Describe an appropriate IBD diagnostic workup
3. Differentiate Crohn’s disease from Ulcerative colitis
4. List available treatment options for IBD

Case
Sarah, a 12-year-old female, comes to your office with a history of bloody diarrhea. Sarah was well until two weeks ago when she started having diffuse abdominal pain, nausea, and loose stools up to five times daily. Her stools became bloody, more frequent and began waking her up during the night. She developed severe urgency and pain with defecation. She also complained of low grade fevers, poor appetite, and fatigue. She denies recent travel or sick contacts.

Her past medical history is negative for any chronic disorders. She has never been hospitalized nor had surgeries in the past. She does not take any medications or herbal supplements. She is not allergic to any medication. Her parents and siblings are healthy, but her maternal grandmother had celiac disease.

Physical examination: WT 32 kg (3%), HT 155 cm (50%-75%), BMI 13 (≤ 3%). She appears pale and tired, but in no acute distress. Her sclera are not icteric and conjunctiva and oropharynx are not hyperemic. She has a small, rounded ulcer on her lower lip. Her neck is without masses or lymphadenopathy. Her lungs are clear to auscultation. Cardiac examination reveals sinus tachycardia without murmurs. Abdomen is soft, not distended, but tender in the right lower quadrant. Her liver and spleen are not enlarged. There are no palpable intraabdominal masses. She has audible bowel sounds. Perianal examination reveals a small skin tag at 9 o’clock. Her skin is well perfused with painful nodules noted on her right shin (see figure 1).

Laboratory Workup:
WBC 5.0 (ref: 4.0-10.5 K/µl), Hemoglobin 9.0 (ref: 12.0-15.0 g/dl), Hematocrit 28 (ref: 35%-45%), and Platelets 384 (ref: 160-370 K/µl) with the following differential 38% neutrophils, 48% lymphocytes, 7% monocytes, 1% basophils. Total protein 7.0 (ref: 6.0-8.0 g/dl), albumin 3.0 (ref: 3.3-4.7 g/dl), calcium 8.4 mg/dl (ref: 8.5-10.2 mg/dl), ESR 40 (ref: 0-20 mm/hr), CRP 4.0 (ref: 0-1 mg/dl), 25-OH vitamin D 20 (ref: 30-80 ng/ml), and vitamin B 12 150 (ref: 210-911 pg/ml).

Stool studies for Salmonella, Shigella, Campylobacter, Yersinia, and Clostridium difficile are negative.

You see Sarah in your office for follow up in one week. Her symptoms are persistent; you refer her to the pediatric gastroenterologist for further IBD workup. An upper endoscopy and colonoscopy are performed (Figures 2 and 3).

Figure 1
Painful nodules on lower extremity
Figure 2. Upper endoscopy and colonoscopy demonstrating a.) erythema and focal erosions in the antrum, b.) aphthous, focal ulcers in the second portion of the duodenum, c.) cobblestoning and deep ulcerations in the sigmoid colon, and d.) loss of architecture, pseudopolyps, deep ulceration and diffuse inflammatory exudates in the transverse colon.

Figure 3. Pathology demonstrating a.) chronic, active colitis with submucosal granulomas and crypt distortion, and b.) a well-organized granuloma in the lamina propria.

Final diagnosis: Based on the clinical presentation, laboratory studies, and histopathology findings, the patient was diagnosed with Crohn’s disease of the small bowel and colon.

Overview of Inflammatory Bowel Disease (IBD)

Inflammatory bowel disease (IBD) is a general term encompassing a range of diseases that cause chronic inflammation in the gastrointestinal tract and are not due to infection or other identifiable causes. The two main types of IBD are Crohn’s disease (CD) and Ulcerative colitis (UC). The Crohn’s and Colitis Foundation of America estimates that as many as one million Americans have either CD or UC. Of these individuals, approximately 20% are diagnosed in childhood. The incidence and prevalence of CD and UC are comparable. Most pediatric epidemiologic studies shown an increased incidence of CD compared to UC; however, in children ages 3-5 years, UC is more common than CD.

To complete this article, read more about the etiology, workup and treatment of IBD in children, and receive CME credit, please go to uwhealth.org/pediatricpathways

Dorota Walkiewicz-Jedrzejczak, MD is a board certified Pediatric Gastroenterologist and an Assistant Professor at the University of Wisconsin School of Medicine and Public Health.

A Letter from the Medical Editor

Dear Colleagues,

In this issue of Pediatric Pathways, we focus our CME component on a range of gastrointestinal complaints encountered while caring for pediatric patients in your office. We provide introductions to these topics in a case-based format in print and give recommendations regarding relevant evaluation, treatment options, and referral information online.

To access this information and receive CME credit, please go to uwhealth.org/pediatricpathways

Please contact me at mkelly@pediatrics.wisc.edu if you have any feedback on this publication or suggestions for future topics.

Sincerely,
Michelle Kelly, MD

Michelle Kelly, MD is a Pediatric Hospitalist and Assistant Professor of Pediatrics at the University of Wisconsin School of Medicine and Public Health.

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On October 10 and 11, 2012, the UW Department of Pediatrics welcomed Holmes Morton, MD, the founder of Pennsylvania’s Clinic for Special Children (www.clinicforspecialchildren.org), and winner of an Albert Schweitzer Prize for Humanitarianism and a MacArthur fellowship, as a visiting professor.

The Clinic for Special Children has cared for children with over 100 rare genetic disorders, advanced newborn screening methods, and furthered clinical research on inherited disorders. Its successes have translated into improved care for all children with inherited disorders—not just those in the Amish and Mennonite communities.

Dr. Morton’s visit focused on improving newborn screening for and health care delivery to Wisconsin’s Amish and Mennonite population—which, at over 16,000 for the Amish and growing rapidly, is the fourth largest in the US.

A ‘Phenomenal’ Community Event
Dr. Morton spent his first day at a community health event in Norwalk, near one of the more concentrated Amish settlements in the state.

Half of the 400 attendees were community practitioners, state and county health personnel, and UW School of Medicine and Public Health (SMPH) faculty, staff, and residents. The remaining half were Amish and Mennonite individuals, families, and midwives—some of whom came from 300 miles away.

Dr. Morton gave an inspiring presentation on ways to improve newborn screening and health care delivery to Wisconsin’s Amish and Mennonite population—which, at over 16,000 for the Amish and growing rapidly, is the fourth largest in the US.

He emphasized that many genetic disorders seen at the clinic can be identified in the first 24 hours of life. What’s more, nearly 80 percent of those disorders are treatable, furthering the case for more comprehensive newborn screening—for all children.

Later, in “Cutting-Edge Diagnostic Testing the Low-Budget Way,” (http://videos.med.wisc.edu/videos/42816), he shared concrete suggestions for advancing newborn screening in Amish and Mennonite populations.

Now, Dr. Seroogy and others are considering how to implement some of his suggestions in Wisconsin. “The one thing [he] hammered home is that you have to be in the community,” Dr. Seroogy said. “There are a lot of babies that are not being seen, and if we had a presence in the community, that could change.”

References
(1) http://www2.etown.edu/amishstudies/Population_by_State_2012.asp

Financial support for the professorship was provided by a grant from the American Academy of Pediatrics and its Wisconsin Chapter (Ellen Wald, MD, principal investigator); the March of Dimes; and the American Family Children’s Hospital.
An 11-year-old Boy with Stunting, Wasting, and Diarrhea

By Peter F. Nichol, MD, PhD

Educational Objectives
After reading this article and answering the review questions, the reader will be able to:

1. Name at least three major causes of pediatric short gut syndrome
2. Differentiate between anatomic and functional short gut syndrome
3. Identify at least one typical nutritional/non-caloric deficit in short gut syndrome
4. Differentiate between short gut syndrome and intestinal failure

Case
James is an 11-year-old boy with a remote history of necrotizing enterocolitis (NEC). He has just moved to your area and presents for a well child check. His family is concerned about enopresis. Upon further questioning he has 3-4 loose stools/day and, when he leaks at night, it is always liquid stool. His history is significant for a recent fracture of his humerus from a fall on pavement from a standing position. His review of systems is also significant for long term general fatigue compared to his peers.

During your exam he complains to his mother that he is hungry, to which his mother remarks, “he is always hungry.” On exam, he is short and very thin with minimal body fat. He has a transverse scar on his abdomen above the umbilicus and another small scar in the right lower quadrant.

Pediatric Short Gut Syndrome
Short gut syndrome is a disease that results from permanent incapacitation of a large portion of the gastrointestinal (GI) tract. The GI tract in this disease is defined as the absorptive regions of the intestine running from the duodenum to the right colon. Shortgut can be segregated into two categories: anatomical disease and functional disease.

In patients with anatomical disease, symptoms arise due to loss of intestinal length and absorptive capacity. In pediatric patients, loss of intestinal length results from congenital defects such as intestinal atresia (Figure 1A), gastrochisis (Figure 1B), and malrotation/volvulus or acquired conditions such as necrotizing enterocolitis (Figure 2) or less frequently Crohn’s disease, traumatic injury to the mesenteric vessels and adhesive bowel obstruction. Symptoms in these patients include: diarrhea, fluid and electrolyte abnormalities, malabsorption, and malnutrition.
The severity of anatomical disease can vary based on the length of intestine that is lost as well as the specific regions that are absent. Accordingly, the intensity of symptoms corresponds to the severity of disease. Thus manifestations of anatomical short gut can range from a child that is hungry all the time and is not growing to a patient that has minimal intestinal length, high GI fluid losses, and is dependent on total parenteral nutrition (TPN).

In functional short gut, the entire intestinal length is present; however, patients are unable to either absorb or tolerate enteral feeds. In children, etiologies include microvillus inclusion disease and the poorly understood entities of pseudo-obstruction and visceral neuropathies. Presenting symptoms in patients with pseudo-obstruction include pain with enteral feeds, nausea, and vomiting whereas patients with microvillus inclusion disease present as newborns with intractable diarrhea. Functional short gut typically does not present with a range of severity. In almost all patients, the disease is severe and incapacitating requiring full nutritional support with TPN.

To complete this article, read more about the management of pediatric short gut syndrome, and receive CME credit, please go to uwhealth.org/pediatricpathways

Peter Nichol, MD, PhD is a board certified Pediatric Surgeon and an Assistant Professor at the University of Wisconsin School of Medicine and Public Health.

Major Expansion Underway at American Family Children’s

Construction is now underway on a 26-bed expansion at the UW’s American Family Children’s Hospital in Madison. Scheduled for completion in early 2014, the project will increase the hospital’s bed count from 61 to 87 and ensure that growing demand for UW pediatric specialists and surgeons will be met for patients and families from throughout the region.

Key highlights of the expansion include:

- A 14-bed Level IV Neonatal Intensive Care Unit (NICU) offering state-of-the-art medical and surgical care partnered with private rooms that give parents the opportunity to stay with and nurture their babies as they begin life. One of only two Level IV NICUs in Wisconsin, this will be the only one with a special emphasis on the newborn requiring surgery and specialized heart and brain monitoring.

- A 12-bed expansion to our Pediatric Intensive Care Unit (PICU), creating highly advanced, private spacious rooms staffed by specialty-trained pediatric critical care physicians, nurses, pharmacists and respiratory therapists.

- Enhancement of our Pediatric Heart Program through construction of a Pediatric Hybrid Cardiac Catheterization/Operating Room/Interventional Radiology Laboratory. This project will ensure that children needing invasive heart and radiological procedures currently performed at the adult UW Hospital will instead have them performed in the Children’s Hospital in a unit specially designed for pediatric care. Only a handful of centers in the Midwest will offer this type of facility.

- Construction of a Pediatric Imaging Suite where children – and only children – will receive X-ray/imaging procedures with state-of-the-art, low-dose radiation technology. This will be a full-service imaging suite for children requiring MRI, CT, ultrasound and fluoroscopy services.

- Opening two more Pediatric Operating Rooms to accommodate the substantial growth in pediatric surgical volume we have seen over the past five years.

“These enhancements will ensure that we have the capacity to care for children and families who need us,” says Donna Katen-Bahensky, president and CEO of UW Hospital and Clinics and American Family Children’s Hospital. “Occupancy has grown substantially since we opened American Family Children’s Hospital in 2007, and we are also seeing more acutely ill patients than most hospitals in Wisconsin that care for children.”

Noting the sense of urgency to complete the project, hospital fundraisers are asking donors to support the “Sick Kids Can’t Wait” Campaign. More information about the campaign – including a special physician society of supporters – is available by visiting uwhealthkids.org

To schedule an appointment with Dr. Nichol in the UW Pediatric Short Gut/Intestinal Failure Clinic or the Pediatric General Surgery Clinic, please call (608) 263-6420 or visit our website at uwhealth.org
A 15-year-old Girl with Chronic Abdominal Pain

By Luther Sigurdsson, MD

Educational Objectives
After reading this article and answering the review questions, the reader will be able to:

1. Define functional abdominal pain
2. Explain the importance of the history and physical exam (rather than extensive investigation) as paramount in the diagnosis of functional abdominal pain
3. Explain the proposed etiology of functional abdominal pain to a patient and parent

Case
Tami is a 15-year-old girl who presents in November to your pediatric GI clinic with the complaint that “her stomach always hurts.” She is accompanied by her parents, who inform you that she is fasting, in case you wanted to perform an endoscopy. The pain has been present for the past ten months, is somewhat better during the summer and does not wake her from sleep. It is described as constant, dull and occasionally crampy, primarily around the umbilicus. There is no association with meals, bowel movements or exercise. She has missed school on six separate occasions this fall and been seen for the pain by her primary care doctor repeatedly and on two occasions in her local ED. Trials of proton pump inhibitors, a stool softener, lactose exclusion and high-fiber diets have not helped.

She has a history of occasional headaches and nausea. No weight loss, mouth ulcers, diarrhea or fever. She has regular menses. She achieves good grades in school, plays soccer, and has not missed games or practices. She denies stressors or negative life events. Family history is significant for migraines but no GI diseases.

Her growth chart and physical exam are normal. Specifically, there is no evidence of abdominal fullness, tenderness or perianal abnormalities. She has had repeated normal workups including CBC, CRP, lipase and liver enzymes along with a normal U/A and pregnancy test. ED visits prompted both a CT and ultrasound of her abdomen, both of which were normal.

The patient and parents’ main concern now is if we are missing something and whether she should try a gluten-free diet.

Definitions and Terminology
Chronic abdominal pain is a common complaint in children. It rarely has an identifiable organic cause but rather is thought to be a functional disorder. The phrase “recurrent abdominal pain (RAP)” has been used since the publication of the seminal article on abdominal pain in children by Apley and Naish in 1958. Most subsequent studies of RAP over the next three decades used their criteria of three or more episodes of pain over three months severe enough to affect the child’s activities. Over time, these entry criteria became a definition and the term recurrent abdominal pain became a synonym for functional abdominal pain (FAP).

The criteria proposed by Apley and Naish in their early papers have been criticized for their ambiguity and for including both organic and nonorganic causes. Recently a subcommittee on chronic abdominal pain from the American Academy of Pediatrics (AAP) and the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) issued clinical and technical reports on chronic abdominal pain recommending abandoning the term recurrent abdominal pain in favor of the term functional gastrointestinal disorder (FGID). The latter phenomenon can be further categorized as functional dyspepsia, irritable bowel symptom, abdominal migraine or functional abdominal pain (see Table 1 online). These terms are defined in the Rome III criteria, a consensus statement proposed by adult and pediatric experts in functional gastrointestinal disorders.

To complete this article and read more about the etiology, workup and treatment of functional abdominal pain in children, and to receive CME credit, please go to uwhealth.org/pediatricpathways.

Luther Sigurdsson, MD, is a board-certified Pediatric Gastroenterologist and an Associate Professor at the University of Wisconsin School of Medicine and Public Health.

To schedule an appointment with Dr. Sigurdsson, please call the UW Pediatric and Gastroenterology Clinic at (608) 263-6420 or visit our website at uwhealth.org
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Questions?
Please contact Barb Anderson, at cme@pediatrics.wisc.edu or (608) 263-8542.

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