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# A Newborn With Abdominal Pain

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A previously healthy 3-week-old boy presented with 5 hours of marked fussiness, abdominal distention, and poor feeding. He was afebrile and well perfused. His examination was remarkable for localized abdominal tenderness and distention. He was referred to the emergency department in which an abdominal radiograph revealed gaseous distention of the bowel with a paucity of gas in the pelvis. Complete blood cell count and urinalysis were unremarkable. His ongoing fussiness and abnormal physical examination prompted consultation with surgery and radiology. Our combined efforts ultimately established an unexpected diagnosis.

## CASE HISTORY

### Dr Riham Alwan (Pediatric Emergency Medicine Fellow) and Dr Meredith Drake (Pediatric Resident):

A 3-week-old boy was referred to the emergency department (ED) from his pediatrician's office with irritability, poor feeding, abdominal tenderness, and distention. In the pediatrician's office he was irritable, and he was noted to have a somewhat rigid, distended, and slightly discolored abdomen in addition to bilateral eye drainage. He was born at 37 weeks and 6 days to a gravida 2 para 2 mother via normal spontaneous vaginal delivery. Cefazolin was administered to his mother predelivery for positive group B *Streptococcus* status. Prenatal, birth, and postnatal course was otherwise unremarkable. Starting at ~2:30 AM on the day of presentation, his parents reported that he had been fussy, with a cry described as "painful." He had been more difficult to console and had been refusing feeds by either breast or bottle. He had 1 episode of forceful emesis 2 days before presentation, and otherwise he had minimal spit up, which was always nonbilious and nonbloody, after feeds. His stools had been soft and seedy, without blood or mucous. He passed meconium within the first 24 hours of life. He had been gaining weight appropriately.

On examination in the ED, he was alert but uncomfortable. His temperature was 37.5°C, pulse was 184 beats per minute, respiratory rate was 40 breaths per minute, and blood pressure was 107/68 mm Hg. The examination of the heart and lungs was normal. He had normal pulses and a capillary refill time of 3 seconds. There were no skin rashes. He had good tone. He had mucous drainage from both eyes without conjunctival injection. His abdomen had positive bowel sounds but was moderately distended with diffuse tenderness after palpation. There was no obvious discoloration of the abdomen. There were no masses or organomegaly. An examination of the genitalia revealed bilaterally descended testes with a normal circumcised penis. The perianal area was normal.

**Dr Matheny, as the physician that initially saw this patient in clinic, please tell us about his initial presentation and examination. What was included in your differential diagnosis? Why did you decide to send him to the emergency department?**

### Dr Cali Matheny (General Pediatrician):

Several of this infant's presenting signs differentiated him from other infants who have fussiness and poor feeding. His symptoms had an abrupt

## abstract

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Dr Alwan initiated this collaborative project, reviewed the literature and interviewed the coauthors, designed the report, and drafted and edited the manuscript; Dr Sobolewski initiated this collaborative project, designed the report, and drafted and edited the manuscript; Dr Drake reviewed the literature and interviewed the coauthors, designed the report, and drafted and edited the manuscript; Dr Emery contributed the ultrasound images and contributed to the drafting, editing, and reviewing of the manuscript; Drs Shaaban and Gurria Juarez contributed the intraoperative images and contributed to the drafting, editing, and reviewing of the manuscript; Dr Szabo contributed the pathology images and to the drafting, editing, and reviewing of the manuscript; and all authors approved the final manuscript as submitted.

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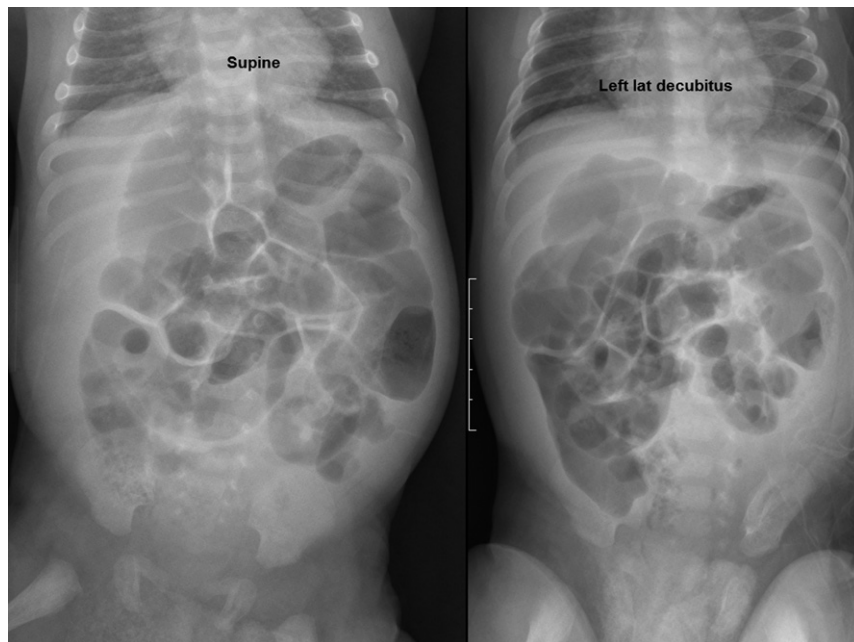
onset. He was irritable and difficult to console. His abdomen was somewhat rigid, slightly distended, and had a mild violaceous hue. I divided my initial differential diagnosis into obstructive and infectious processes, and I believed an obstructive process was more likely given his feeding refusal and the marked decrease in his usual stooling pattern. I was concerned for malrotation with volvulus, intussusception, necrotizing enterocolitis (NEC) and short-segment Hirschsprung disease. I believed that constipation was unlikely given that he was exclusively breastfed. Potential infectious processes included serious bacterial infection (eg, a urinary tract infection). His abdominal examination also made me concerned for NEC. Given this differential, I referred him to the emergency department for further evaluation and treatment including radiographic studies that were not available in my office.

**Drs Alwan and Drake:**

**Dr Sobolewski, as a pediatric emergency medicine attending, please share your thoughts about this patient's initial differential diagnosis given the chief complaint and presenting symptoms of fussiness, abdominal pain, and distention? What would be the initial steps in the diagnosis process?**

**Dr Brad Sobolewski (Pediatric Emergency Medicine):**

My initial differential diagnosis aligned with Dr Matheny's and included bowel obstruction (small bowel or distal [eg, Hirschsprung disease]), malrotation with volvulus, neonatal sepsis, NEC, urinary tract infection, constipation, hypertrophic pyloric stenosis, infantile colic, gastroesophageal reflux, nonaccidental trauma, and gastroenteritis. Given that he was consolable, afebrile, and his fussiness seemed to localize to the abdomen, we felt that eschewing the traditional



**FIGURE 1**

Abdominal plain radiographs showing mild nonspecific gaseous distension without pneumoperitoneum.

sepsis workup and focusing on the abdomen first was prudent. The initial evaluation therefore began with an abdominal radiograph, which can demonstrate signs of bowel obstruction.

His abdominal radiograph showed diffuse gaseous distension of the bowel with a paucity of gas in the pelvis (Fig 1).

**Drs Alwan and Drake:**

**Dr Emery, as the radiologist who read this abdominal film, what do these findings suggest to you? What diseases can present with these imaging findings in a neonate? Are there other images that may be helpful to obtain initially?**

**Dr Kathleen Emery (Pediatric Radiology):**

The findings in this patient's abdominal radiograph are highly nonspecific. Lack of gas in the pelvis could indicate a distal obstruction or the dependent position of the rectosigmoid. Supine and decubitus are the 2 most useful views to obtain initially. I do not recommend prone

views unless there is significant clinical concern for distal obstruction. In that case, then I would recommend placing the neonate on his abdomen for few minutes and evaluating with a prone view for persistent paucity of gas.

Given the history, this patient could be suffering from early NEC, intussusception, malrotation with volvulus, Hirschsprung's disease, or more commonly, gastroenteritis. If the patient had bilious emesis, I would have suggested an upper gastrointestinal study. Ultimately the history was as nonspecific as the abdominal radiograph.

**Drs Alwan and Drake:**

After reviewing the radiograph we performed a rectal examination, as this radiograph finding could suggest Hirschsprung's. After the insertion of a finger into the rectum the patient expelled copious watery green stool. His fussiness was only relieved temporarily, and he continued to refuse oral intake. Because the rectal examination was concerning

for distal obstruction, namely Hirschsprung disease, surgery was consulted.

**Drs Shaaban and Gurria Juarez, what findings on initial history and physical examination concern you? What other historical elements are more consistent with Hirschsprung disease and other causes of bowel obstruction in neonates? What specifically in this patient made you question the possible diagnosis of Hirschsprung disease? After evaluating the patient, what was your plan?**

**Dr Aimen Shaaban (Pediatric Surgery Attending) and Dr Juan Gurria Juarez (Pediatric Surgery Fellow):**

On initial evaluation in the ED, this patient was clinically stable but irritable. He had a distended but soft, mildly tender abdomen with normoactive bowel sounds. Rectal stimulation was performed with a feeding catheter with immediate return of gas and stool. The history of present illness guided our discussion with the parents toward the possibility of gastroenteritis. We felt that Hirschsprung disease was less likely because he had been healthy and asymptomatic during his initial weeks of life, with a history of passing meconium within the first day of life and having no difficulties stooling. His acute deterioration in health therefore lessened concerns for Hirschsprung disease. The spectrum of Hirschsprung disease encompasses different clinical presentations that can vary from mild constipation to fulminant toxic enterocolitis.<sup>1</sup> It commonly presents with failure to pass meconium within the first 24 hours of life and progresses to repeated episodes of constipation.<sup>2</sup> In cases that progress to enterocolitis, infants typically develop tense abdominal distention over several hours and have profuse vomiting along with large amounts of foul smelling gas

and stool.<sup>1,2</sup> Although our patient exhibited abdominal distention, the rest of his clinical picture and history did not fit with the diagnosis. Radiographic findings in our patient were rather nonspecific, and along with his physical examination, the decision was made to treat him nonoperatively. Should his symptoms persist despite medical management, we would then evaluate for Hirschsprung disease.

The differential diagnosis of bowel obstruction in neonates is broad and diverse depending on the age of the patient. It involves the anatomy from mouth to anus, including esophageal webs, atresia, pyloric stenosis, duodenal web and/or duodenal atresia, annular pancreas, malrotation, jejunoileal atresia, duplications, meconium ileus, microcolon, constipation, Hirschsprung disease, left colon syndrome, atresia, and imperforate anus, among others.

**Drs Alwan and Drake:**

After discussing the patient with our surgical colleagues, the patient's mom attempted to breastfeed the patient. He would latch onto the breast for only a few seconds and then pull away. He similarly refused the bottle. At this point, we ordered a complete blood cell count (CBC) and blood culture, catheter urinalysis and urine culture, a basic metabolic panel and a 20 mL/kg normal saline bolus. If the results of the CBC were worrisome, we discussed the pursuit of a lumbar puncture to evaluate for meningitis.

Ultimately, the results of the CBC, electrolytes, and urinalysis were unremarkable. This left us in a bit of a diagnostic quandary, and we were prepared to admit to general pediatrics.

**Dr Sobolewski:**

What unsettled me was that although the patient fell asleep after the

laboratories had been obtained, his abdominal examination continued to be impressive for localized tenderness to the lower abdomen on repeat examination 2 hours after arrival. In fact, I felt that he had outright peritonitis with involuntary guarding and that his acute abdomen would best be evaluated by obtaining an ultrasound.

**Drs Shaaban and Gurria Juarez, what can you tell us about the term infant with peritonitis? What does it suggest in terms of the differential diagnosis and how do you best evaluate it?**

**Drs Shaaban and Gurria Juarez:**

The cause of peritonitis in this population is often because of an acute obstruction or perforation as a complication of NEC or intussusception. NEC typically affects preterm infants and presents with abdominal distention and vomiting. As their abdominal distention worsens they may develop occult positive or grossly bloody stools. The abdominal wall may appear erythematous. Abdominal radiographs are used to make the diagnosis and reveal pneumatosis intestinalis or intramural gas.<sup>1</sup> Although the initial examination by the primary medical doctor included abdomen discoloration and distention, our patient was born almost to term, had nonbloody stools, and did not have any of the findings on radiograph that are consistent with NEC.

The evaluation usually begins with blood gas, CBC with differential, C-reactive protein and/or erythrocyte sedimentation rate, lactate level, electrolytes, and blood cultures as well as adjuncts to physical examination such as plain radiographs and ultrasound.<sup>3</sup> However, an urgent surgical exploration should be considered early in the evaluation of an infant with peritonitis because catastrophic diseases such as midgut volvulus are best diagnosed in the operating room

during laparotomy such that the time interval to treatment is minimized. Unfortunately, this requires acting based on a limited amount of data. Waiting for laboratories could have delayed further care and would not have been diagnostic. The lack of bilious emesis in this case diminished the likelihood of midgut volvulus and the more indolent presentation provided support for a limited workup. Fortunately, we were able to obtain an ultrasound quickly, and ultrasound results made the additional laboratory tests unnecessary.

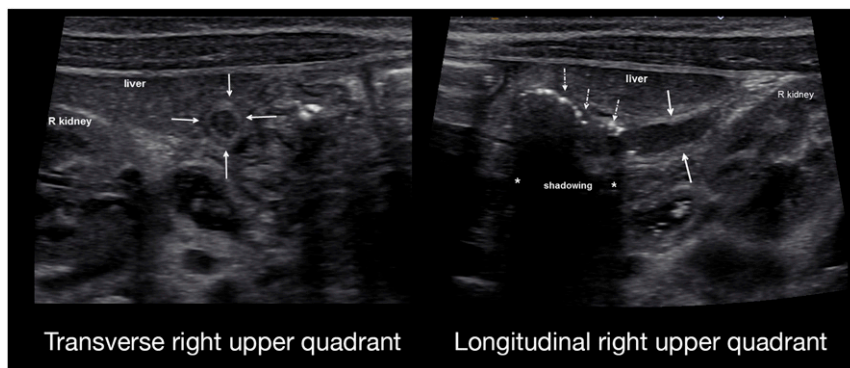
**Dr Sobolewski:**

**Dr Emery, is there anything unique about reading an ultrasound in the newborn? What are some of the limitations of ultrasound in this age group? Why is it a better choice than computed tomography scan?**

**Dr Emery:**

Appropriately, you picked up the phone and called me saying, “There is something funny about this kid. He has abdominal tenderness of some etiology.” This speaks volumes to me. If you are worried, communicate your concerns directly to the radiologist. Infants can have significant amounts of bowel gas that can limit the utility of the ultrasound. Their anatomy is small, but usually the ultrasound is an excellent adjunct to the physical examination, if the abdominal radiograph has been unrevealing.

After speaking to the clinical team, my differential diagnosis still included NEC and intussusception. I have definitely seen both in term infants. Gastroenteritis is common. Anatomic abnormalities such as the malrotation spectrum can also present this way. I think in an infant that young without fat planes that a computed tomography scan of the abdomen without oral contrast is significantly limited. The ultrasound is a better imaging



**FIGURE 2**

Transverse ultrasound showing the dilated appendix (arrows) measuring 6 mm in diameter located adjacent to the inferior edge of the liver. Longitudinal image demonstrating a tubular structure (solid arrows) with hypoechoic fluid in the lumen near the right lobe of the liver and right kidney. The punctate echogenic foci in the wall (cashed arrows) are consistent with air (asterisks).

modality. There is some literature to suggest ultrasound can be useful in detecting pneumoperitoneum, but many radiologists still recommend initial abdominal radiographs for the evaluation of NEC.<sup>4</sup> According to Schwartz et al,<sup>3</sup> in an infant with signs of abdominal sepsis, the next step after obtaining an abdominal radiograph that is negative for signs of NEC is to obtain an ultrasound of the intestines.<sup>5</sup>

On ultrasound, the appendix is usually found after first localizing the cecum. This case was difficult because the appendix was nestled in the right upper quadrant near the inferior edge of the liver, adjacent to the lower pole of the kidney (Fig 2). One of the advantages to ultrasound is the dynamic element during imaging, which improves its utility in an age group that is otherwise unreliable on physical examination.<sup>3</sup> This child, for example, cried immediately with attempts to compress his appendix. The ultrasound revealed an appendix that was enlarged and fluid filled. There appeared to be air within the appendiceal wall and impending perforation was an acute concern (Fig 2). In my 27 years, I have never seen a case like this. I see signs of ischemia or localized NEC far more often. Air in the appendiceal wall is extremely rare.

**Drs Alwan, Drake and Sobolewski:**

Because the ultrasound was diagnostic for acute appendicitis, we asked surgery to urgently re-evaluate the patient.

**DISCUSSION**

**Final Diagnosis: Acute Appendicitis**

**Dr Matheny:**

I think that the important message from this case is that parental calls regarding fussy infants <2 to 3 months of age should be taken seriously. Young infants with fussiness should generally be seen the same day. If the office is closed, these patients should be referred to an ED as opposed to an urgent care. Although the majority of these infants will have common diagnoses, such as developmentally typical crying or colic, some will have more serious illnesses, such as urinary tract infections. Freedman et al<sup>6</sup> reviewed 237 afebrile infants <1 year of age who presented to a pediatric ED with crying or fussiness, and they found that ~5% had a serious illness. If the infant is ill, and especially if they have an acute abdomen, they should be promptly referred to an ED whether diagnostic testing such as radiographs and urinalysis are available in the office.

**Drs Alwan, Drake and Sobolewski:**

**Drs Shabaan and Gurria Juarez, how rare is appendicitis in the newborn? How does its presentation differ from that of an older child? What concerns do you have operating on a neonate with this diagnosis? Did you experience any unforeseen challenges intraoperatively? During the operation, were the ultrasound findings consistent with the intraoperative findings? How was this like NEC or other intraabdominal calamities in the newborn period?**

**Drs Shaaban and Gurria Juarez:**

Appendicitis in infancy is extremely rare but can occur at any age even in premature infants.<sup>7-9</sup> Hence, the wisdom of the surgical dictum, “appendicitis should never be lower than number 3 on the list of differential diagnoses for an acute abdomen.” Clinical findings of appendicitis in the newborn are nonspecific and universally reflect peritonitis from perforation. A high index of suspicion is unlikely given the rarity of this disease, but a surgical bias toward intervention in the infant with a clinical examination suggesting peritonitis provides a critical safety net to prevent missing such a rare diagnosis.

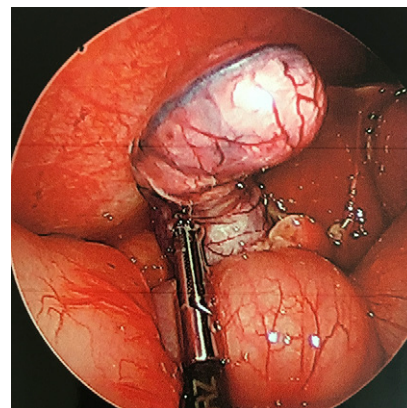
Given the overwhelming concern for a surgically-relevant intraabdominal pathology, the decision to perform a diagnostic laparoscopy was straightforward. In these cases, converting to an open laparotomy is frequently necessary for both diagnostic and therapeutic considerations.<sup>7</sup> The technical approach to appendectomy in a newborn is similar to older children except that the smaller size precludes the use of surgical staplers.

In our patient, the appendix was visualized through an umbilical laparoscopic port to be located in the right upper quadrant. Fortunately, the length of the appendix was

free from attachment to the retroperitoneum making it amenable to externalization via the incision for the second laparoscopic port placed in the right upper quadrant (Fig 3). After externalization, the appendectomy was easily completed by using conventional techniques. In general, the ultrasound accurately predicted the intraoperative findings. However, we found also signs of perforation with purulent fluid throughout the entire abdominal cavity. Thus, copious lavage of the peritoneal cavity was performed before closure. There were no other relevant findings during the exploration.

**Drs Alwan and Drake:**

In reviewing the literature, childhood appendicitis is well described, but neonatal appendicitis is much rarer, with an incidence of 0.04% to 0.2%.<sup>3-5,9-12</sup> There have been ~50 cases reported in the past 30 years, most commonly involving premature boys.<sup>7,9,11</sup> Perhaps given the insidious onset of this disease and the difficulty of the neonatal examination, the mortality rate has been reported as 20% to 25%.<sup>1,3,7-10</sup> Some speculate that the broadness of the appendix, or its conical shape, combined with the lack of fecaliths in neonates contributes to the rarity of this diagnosis.<sup>7,9,12-14</sup> Anatomically, the neonate’s appendiceal wall is thinner and perhaps more easily perforated.<sup>7,9,15,16</sup> The high morbidity and mortality rate could be attributed to faster progression to perforation, peritonitis, and subsequently septic shock.<sup>3</sup> It is important to evaluate each infant on the basis of their unique presentation, and keep neonatal appendicitis on the differential for abdominal distension and feeding difficulty. Of note, Hirschsprung disease and NEC should always be considered when contemplating neonatal appendicitis. Schwartz



**FIGURE 3**  
Intraoperative view of the appendix.

et al<sup>3</sup> present a proposed visual algorithm for neonates with abdominal sepsis as a useful aid.

**Dr Szabo, is your analysis of the appendiceal specimen consistent with the diagnosis or could this be a rare form of localized NEC? Did the histology differ from appendicitis in older children?**

**Dr Sara Szabo (Pathology):**

It is not entirely classic for acute appendicitis, but here is what I can tell you based purely on histology. It is diagnostic of an advanced case of appendicitis. It did not appear to be consistent with more extensive bowel disease as seen in NEC. The pathology report reinforces the intraoperative findings consistent with acute gangrenous appendicitis. Sections of the appendix showed extensive and near complete effacement with destruction of the mucosa and muscularis propria by an active chronic inflammatory infiltrate containing neutrophils with lymphocytes, rare plasma cells, and eosinophils (Fig 4). It is essentially obliterated by what appears to be ischemic coagulative necrosis. There was no fibrinopurulent serositis, which would have developed had the inflammation resulted from an adjacent intraabdominal process, such as an abscess. If this child were younger, you could insinuate that

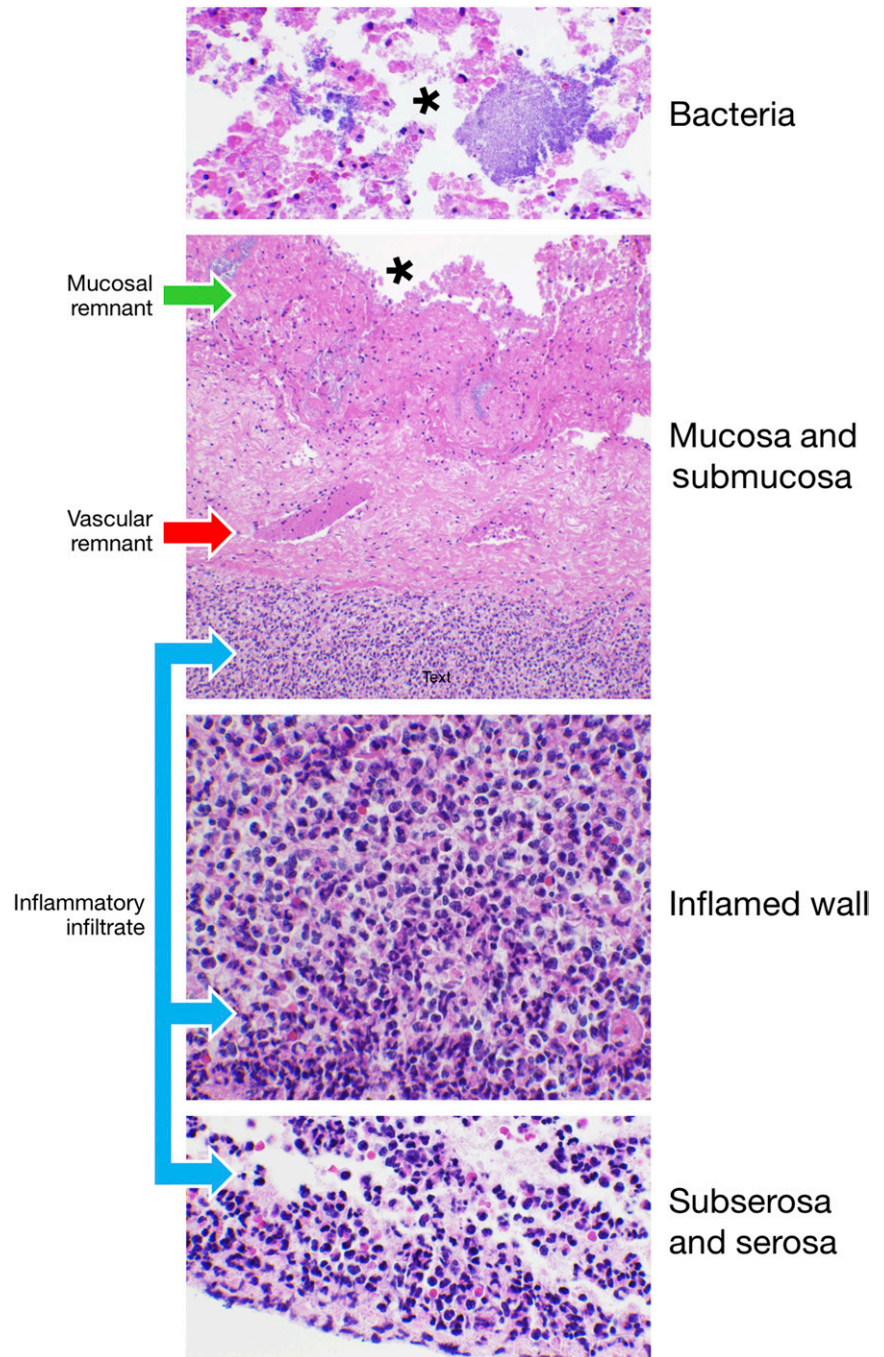
meconium obstructed the lumen, not unlike how a fecalith would obstruct the lumen in an older child. However, his gut appears to be colonized, and the lumen was only slightly dilated. Although obstruction secondary to a Ladd's band, gut malrotation, or incarceration from a hernia had entered the differential, these etiologies have been excluded based on the surgeon's intraoperative findings.

**Drs Alwan, Drake and Sobolewski:**

**Drs Shabaan and Gurria Juarez, was his postoperative course prolonged or complicated? How is the neonatal post-op course different from older children?**

**Drs Shaaban and Gurria Juarez:**

The major differences that exist between infants and older children or adults relate to the well-documented higher rate of perforation and sepsis.<sup>7</sup> The infant or toddler is often perforated (or microperforated or gangrenous) at the time of presentation.<sup>5,7,9</sup> This is because of a delay in diagnosis and treatments as well as the fact that the appendiceal wall is thinner in neonates and they have a relatively indistensible cecum. As a consequence, the postoperative course is that of a complicated appendicitis. Additionally, the infant tends not to form a loculated or contained abscess but more often has a free perforation with overt sepsis. The omentum is almost nonexistent thus the contamination will disperse faster. This explains the higher rates of peritonitis and mortality in neonates compared with older children and adults. Although the patient had signs of perforation in the operating room, he had an uneventful postoperative course. He achieved full feeds by post-op day 2 and was discharged from the hospital on post-op day 3. He was found to have a small incisional seroma during his follow-up clinic visit, which resolved with time.



**FIGURE 4** Pathology specimen showing acute necrotizing appendicitis. Top to bottom. Bacteria are seen throughout the lumen of the specimen. The mucosa is entirely devitalized (green arrow), with bacterial colonies growing on sloughed luminal debris (asterisk). The submucosa is largely ischemic, with vascular remnants (red arrow). A heavy active inflammatory infiltrate (blue arrows) has overrun the entire muscular wall and subserosa, with remnant ischemic and reactive smooth muscle fibers.

**Dr Matheny, how is the patient doing now?**

**Dr Matheny:**

He developed diaper dermatitis shortly after discharge, which was likely

secondary to diarrhea from Augmentin use. He has had multiple well child visits since that time and has continued to thrive developmentally and have normal growth, and he has had no issues after surgery.

## CONCLUSIONS

The initial evaluation of the fussy neonate is uniquely challenging. An appropriately thorough history and careful physical examination guides development of the differential diagnosis. In newborns with anorexia and localized abdominal tenderness consider an initial radiograph followed by ultrasound.

Appendicitis should always be considered despite its relative rarity. Ultimately, our patient was correctly diagnosed with acute appendicitis and subsequently recovered because of the high index of suspicion. Communication and collaboration among physicians, surgeons, radiologists, and pathologists allowed for an accurate and timely diagnosis.

## ACKNOWLEDGMENT

We thank Dr Cali Matheny for her essential input and advice on this case.

## ABBREVIATIONS

CBC: complete blood cell count  
ED: emergency department  
NEC: necrotizing enterocolitis

**COMPANION PAPER:** A companion to this article can be found online at [www.pediatrics.org/cgi/doi/10.1542/peds.2017-0568](http://www.pediatrics.org/cgi/doi/10.1542/peds.2017-0568).

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