Case 1: Acute, Intermittent, and Colicky Abdominal Pain in a 12-year-old Boy
Case 2: Emesis Since Birth in a 2½-month-old Infant
Case 3: Worsening Cough Since Birth in a 16-month-old Toddler

Case 1 Presentation
A 12-year-old boy presents to the emergency department with acute-onset, intermittent, colicky right lower quadrant abdominal pain for 3 to 4 hours. The pain is associated with nausea and one episode of non-bilious, nonbloody vomiting. He has not had diarrhea, constipation, fever, urinary symptoms, trauma, or past similar episodes.

On examination, the boy’s temperature is 38.8°C, his heart rate is 146 beats per minute, his respiratory rate is 26 breaths per minute, and his blood pressure is 110/65 mm Hg. He looks tired. His weight is 38.8 kg (35th percentile). Abdominal examination reveals a soft and nondistended abdomen, but he is tender in the left and right lower quadrants. He has no rebound tenderness, guarding, palpable masses, or organomegaly. The findings on the rest of the physical examination are unremarkable.

Initial laboratory evaluations show hemoglobin of 13.6 g/dL, a white blood cell count of $18 \times 10^3/\mu L$ (82% neutrophils, 12% lymphocytes, 6% monocytes, and 1% eosinophils), and a platelet count of $277 \times 10^3/\mu L$. His blood levels are as follows: total serum bilirubin, 0.6 mg/dL; direct bilirubin, <0.1 mg/dL; alanine aminotransferase, 18 IU/L; aspartate aminotransferase, 28 IU/L; alkaline phosphatase, 175 IU/L; amylase, 34 IU/L; and lipase, 42 IU/L. Serum electrolyte levels are normal. The only abnormality on urinalysis is 1+ blood. Abdominal radiographs show a nonspecific bowel gas pattern. Ultrasonography of the abdomen reveals elongated tubular structures with hypoechoic centers in the small intestine, which suggest a diagnosis (Fig 1). An additional study confirms the diagnosis.

Case 2 Presentation
A 2½-month-old boy is referred to our hospitalist service from an outside hospital. His parents report daily, nonbloody, nonbilious vomiting since birth. The vomiting often is worse after feeding and has been so frequent that the patient has not gained weight appropriately. His stools have been normal. In addition, he has had frequent episodes of screaming and arching of his back each day that last several hours and occasionally are associated with feeding. His prenatal history was unremarkable, and his birthweight was at the 50th percentile. A gastroenterologist has diagnosed the infant as having milk allergy and gastroesophageal reflux, but trials of hydrolyzed formulas and lansoprazole have not improved his symptoms.

On the day of transfer, the patient is smiling and interactive. His weight is at the 5th percentile, his head circumference is at the 5th percentile, and his vital signs are normal. Findings on examination are completely normal, including his abdominal and neurologic evaluations. A basic metabolic panel on the day of transfer reveals the following levels: sodium, 136 mEq/L; potassium, 3.3 mEq/L; chloride, 102 mEq/L; bicarbonate, 24 mEq/L; BUN, 12 mg/dL; creatinine, 0.4 mg/dL; and glucose, 93 mg/dL.

The evaluation performed at the outside hospital included MRI of the
The clinical presentation depends on the load of infestation and the organs involved. Most children with a low to moderate load remain asymptomatic. Some develop vague abdominal symptoms and may show signs of malnutrition. The most common acute presentations are due to obstruction of the intestine or biliary tract and pulmonary symptoms. Loeffler pneumonia occurs during the passage of larvae through the lungs and is characterized by coughing, wheezing, and difficulty of breathing, associated with blood eosinophilia with fleeting pulmonary infiltrates. More serious conditions, including acute appendicitis, toxic megacolon, liver abscess, ascending cholangitis, and pancreatitis, can occur rarely.

Diagnosis
Traditionally, the demonstration of fertilized or unfertilized eggs in stool has been diagnostic; however, high-density, higher frequency ultrasonography of the abdomen sometimes can be useful in establishing the diagnosis. The live worm on longitudinal section appears as a writhing tubular shadow having brighter margins, described by some as the “strip sign.” The hypoechoic core produces the “inner tube sign.” The cross-sectional picture is also characteristic of a tubular body, described as the “ring sign” or “bull’s-eye sign” (Fig 2).

Treatment and Prevention
Treatment consists of antihelminthic medications, including albendazole in a single dose, mebendazole for 3
days or as a single dose, and ivermectin in a single dose. A follow-up stool specimen at 3 weeks after treatment to assess elimination of eggs can be done but is not essential. Intestinal obstruction as a complication of ascariasis primarily requires conservative management. Piperazine may have a role in the treatment of intestinal obstruction but is not available in the United States. However, surgical interventions may be required in certain situations, such as intestinal obstruction with failed conservative management, complete intestinal obstruction, biliary obstruction, volvulus, or perforation peritonitis.

Loeffler pneumonia primarily requires symptomatic treatment, including bronchodilators, oxygen, and corticosteroids. Some clinicians recommend postponement of anthelmintic treatment until the patient is clinically stable because migration of worms after the anthelmintic treatment may worsen the symptoms. Better sanitation and personal hygiene are important in prevention.

This patient was treated with a single 500-mg dose of mebendazole, was observed overnight, and was discharged after the pain resolved.

Lessons for the Clinician
- Helminthic infestation should be considered as a cause of abdominal pain, especially in immigrant populations.
- Ascariasis usually presents with mild abdominal symptoms. However, occasionally, the infestation may cause serious complications.
- Abdominal ultrasonography may be helpful in the diagnosis of ascariasis.

Case 2 Discussion

Complete abdominal ultrasonography performed to re-evaluate the pylorus and search for other potential causes for the boy’s symptoms demonstrated grade 3 hydronephrosis on the left with no evidence of hydrourerter. A voiding cystourethrogram revealed a normal bladder without vesicoureteral reflux. A 99mTc-mercaptoacetyltriglycine (MAG3) renal scan demonstrated equal renal function, with prolonged uptake of tracer and delayed washout on the left.

These findings were suggestive of left ureteropelvic junction (UPJ) obstruction. The patient underwent a left pyeloplasty, with excision of the narrowed UPJ segment and placement of an internal ureteral stent. The next day, he tolerated feeds without difficulty. The parents reported that he was finally a “happy baby.” Follow-up visits have shown that his vomiting has resolved, and he has been gaining weight well. On repeat ultrasonography, he has mild residual hydronephrosis on the left. Repeat MAG3 renal scan demonstrates normalization of renal drainage with preservation of function.

Case 2

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Figure 2. Abdominal ultrasonography showing a cross-section of a tubular structure, which is a roundworm.

Differential Diagnosis

The differential diagnosis of an infant who presents with nonbilious vomiting depends largely on the duration of the vomiting. When vomiting has been present since birth, overfeeding, gastroesophageal reflux, proximal intestinal obstruction, metabolic disorders, and increased intracranial pressure should be considered strongly. Some disorders causing chronic vomiting in infancy are related to allergy, such as protein-induced eosinophilic gastropathies. Therefore, it is critical to take a thorough history regarding diet, relationship of the vomiting to eating, and presence of diarrhea or hematochezia.

The infant’s growth and development also can provide clues to diagnosis. Developmental delay might suggest a metabolic disorder or increased intracranial pressure. Failure to thrive could be due to the degree of vomiting and calorie loss or could be secondary to an underlying condition. The physical examination should include the infant’s state of hydration, as well as thorough neurologic, abdominal, and genital assessments.

Further diagnostic evaluation of an infant who experiences chronic vomiting should be based on the diagnoses considered most likely after a careful history and physical examination are performed. A complete metabolic profile can offer clues to the diagnosis or can indicate the severity or chronicity of the vomiting. Hyperchloremic metabolic alkalosis suggests that the vomiting has been severe, and hypoglycemia, metabolic acidosis, or elevated liver enzyme levels might suggest a metabolic disorder.

If vomiting is related to feedings, gastroesophageal reflux and proximal intestinal obstruction should be considered, and an upper GI series, pH probe testing, or abdominal ultrasonography should be performed.
If developmental delay or abnormal results on neurologic examination are noted, imaging of the brain or a metabolic evaluation might be indicated. If an allergic condition is suspected, elimination of intact milk protein from the diet and the use of elemental formula should be undertaken, and esophagogastroduodenoscopy can be performed to look for evidence of inflammation or tissue eosinophilia.

In this patient, an elimination diet did not improve the symptoms, and the results of biopsies showed no evidence of allergy. These findings suggest that milk allergy was the incorrect diagnosis. The discovery of the unilateral hydronephrosis led to the evaluation for UPJ obstruction. In hindsight, the mother remembered that she had been told that prenatal ultrasonography had indicated that one kidney was “slightly larger” than the other, but that the difference was not significant.

The Condition
UPJ obstruction is the most common cause of fetal hydronephrosis, accounting for approximately 50% of such cases, and the finding is more common in males. Causes for UPJ obstruction include fibrosis, an aperistaltic segment, or compression by a renal vessel. Two thirds of all cases occur on the left side, and 10% to 40% of cases are bilateral. The incidence ranges from 1 in 500 to 1 in 1,500. Some affected children present later in childhood.

Most neonates and young children afflicted with the disorder are asymptomatic, whereas older patients frequently have symptoms. Acute obstruction and subsequent distention of the renal pelvis activate parasympathetic afferent fibers, causing nausea, vomiting, or pain. Some patients, including those with intermittent obstruction, have impaired drainage of urine and therefore develop symptoms only when urine production exceeds the drainage of urine from the renal pelvis. These patients often will note flank pain and nausea after periods of increased fluid consumption, a phenomenon known as Dietl’s crisis.

The strictest definition of UPJ obstruction includes delayed drainage in combination with either symptoms or impaired renal function. The most common test used in the diagnosis is a nuclear renal scan such as MAG3. This type of scan can assess the drainage of each kidney and can provide information on the contribution of each kidney to overall renal function.

Affected children who show no evidence of impaired renal function may be observed over time with routine ultrasonographic follow-up, because in 50% to 75% of cases, UPJ obstruction will resolve spontaneously without the patient developing any symptoms. However, if renal function is impaired or if symptoms develop, corrective surgery should be considered. The most common surgical technique is an open or laparoscopic pyeloplasty, which involves excising the obstructing segment and re-anastomosing the ureter to the renal pelvis. Complications of pyeloplasty include extravasation of urine and recurrence of obstruction due to scar formation. A few children with impaired renal function before surgery may experience declining renal function despite correction of the obstruction.

Lessons for the Clinician
• Ureteropelvic junction (UPJ) obstruction is a rare cause of vomiting and pain in infants.
• Gastrointestinal (GI) pathology is a much more common cause of vomiting in infants and should be evaluated fully and treated before considering other causes.
• Failure to identify GI causes should prompt an evaluation for other disorders.

Case 3 Discussion
The chest radiograph revealed hyperlucent, gas-filled areas in the right lower chest, suggesting a diaphragmatic hernia. A lateral chest radiograph confirmed the diagnosis of diaphragmatic hernia (Figs 3 and 4). Computed tomography scan of the chest confirmed an anterior diaphragmatic defect, also known as a Morgagni hernia (Fig 5). The diaphragmatic hernia was repaired electively.

The Condition
In the developing fetus, the septum transversum, bilateral pleuropertitoneal membranes, chest wall muscles, and dorsal mesentery of the esophagus contribute to the formation of the diaphragm. During week 8 of gestation, these structures fuse, and thus the pleural and peritoneal cavities are separated. The fusion of the septum transversum and pleuropertitoneal membranes results in the pleuropertitoneal canal. The failure of closure of the pleuropertitoneal canal during gestational weeks 8 to 10 results in congenital diaphragmatic hernia (CDH).

The incidence of CDH is 1 in 2,000 to 3,000 live births. CDHs are classified traditionally as Bochdalek (posterolateral), Morgagni (anterior), and hiatal hernias. Approximately 85% are left-sided Bochdalek hernias.

In classic posterolateral CDH, herniation of intraabdominal organs into the thoracic cavity causes both
ipsilateral and contralateral pulmonary hypoplasia and associated pulmonary hypertension. The compromised developing lung in a fetus with CDH is surfactant deficient and associated with anatomic changes that predispose to pulmonary hypertension. In contrast, most anterior Morgagni hernias are not associated with these pulmonary findings, and incidental discovery is more common.

Approximately one half of the cases are diagnosed in the prenatal period by using ultrasonography. However, prenatal CDH diagnosis rates are variable, depending on the incidence of prenatal screening with ultrasonography.

Infants presenting with symptomatic CDH typically have acute respiratory compromise that may require prompt airway control and assisted mechanical ventilation and oxygenation. An orogastric tube should be placed early to prevent gastric distention and further pulmonary compromise. The diagnosis is confirmed with the use of chest radiographs. Echocardiography may be useful to determine whether congenital cardiac lesions exist and to quantify the degree of pulmonary hypertension.

Some infants with CDH will present with subtle nonspecific signs and symptoms such as recurrent cough and repeated respiratory infections, bouts of variable respiratory distress, and failure to thrive. Morgagni hernias tend to be less symptomatic than posterolateral hernias, and a high degree of suspicion, along with a careful history, examination, and diagnostic index of suspicion

Figure 3. Radiograph of the chest shows intestines in the lower lung areas on both sides and also overriding the cardiac image.

Figure 4. Lateral radiograph of chest showing intestines in the retrosternal area.

Figure 5. Computed tomography scan of the chest showing intestines within the thorax.
imaging, is required. Clinicians should realize that a previously normal chest radiograph does not completely exclude a diagnosis of CDH.

Management

The treatment of CDH is individualized, based on anatomic location, physiologic status, age, and associated congenital anomalies. Posterolateral CDH in a newborn may be acutely symptomatic, requiring escalation of ventilatory management to provide adequate gas exchange. This therapy includes the use of high-frequency oscillatory ventilation, inhaled nitric oxide, and extracorporeal membrane oxygenation support. Therefore, newborns born with posterolateral CDH experience considerable morbidity and mortality reflective of the anatomic and physiologic derangement.

In contrast, most anterior Morgagni hernias do not present with pulmonary hypoplasia and pulmonary hypertension. Therefore, the diagnosis typically is delayed for months to years, and the condition may be found incidentally. After accurate diagnosis of a Morgagni hernia, prompt elective operative repair is indicated to prevent respiratory distress and potential GI obstruction from incarceration. The expected outcome should be excellent, with low morbidity and near-zero mortality.

Lessons for the Clinician

- Congenital diaphragmatic hernia (CDH) presents with a spectrum of signs and symptoms based on anatomic location and physiology.
- Unlike posterolateral CDH, presenting symptoms may be subtle and diagnosis delayed in those born with anterior Morgagni hernias.

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Clarification

In the “Care of the Well Newborn” article in the January 2012 issue (Warren JB, Phillipi CA. Pediatr Rev. 2012;33(1):4–18), erythromycin eye ointment is cited as the most commonly used medication to prevent ophthalmia neonatorum. Although it is effective in the prevention of gonococcal ophthalmia, erythromycin ointment is not effective in preventing conjunctivitis caused by Chlamydia trachomatis. Optimal prevention of ophthalmia involves screening and treating pregnant women for both infections. Ointment administration should not promote complacency in clinicians, who should thoroughly evaluate conjunctivitis appearing in the first month after birth, especially when the mother has not been screened for Chlamydia infection.
Index of Suspicion
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