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STATEMENT OF FINANCIAL DISCLOSURE

To reveal any potential bias in this publication, and in accordance with Accreditation Council for Continuing Medical Education guidelines, we disclose that Dr. Dietrich (editor), Dr. Skrainka (CME question reviewer), Dr. Stanich (author), Dr. Bellolio (author), Dr. Taylor (peer reviewer), Ms. Coplin (executive editor), and Mr. Springston (associate managing editor) report no relationships with companies related to the field of study covered by this CME activity.

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Intussusception and Midgut Volvulus

Abdominal pain is a common chief complaint in the pediatric emergency department (ED) and can be a diagnostic dilemma, as clinical presentation varies greatly depending on the child's ability to communicate.^{1,2,3} In addition, the etiology of abdominal pain differs depending on the child's age and so the differential diagnosis will vary.^{4,5} It is imperative for ED physicians to differentiate those with life-threatening pathology requiring immediate intervention from those with self-limiting causes of abdominal pain. Fortunately, most cases are non-threatening and many children will be discharged home with minimal intervention. However, those with abdominal catastrophes require prompt diagnosis, as delays in diagnosis can prove to be fatal.

— Ann M. Dietrich, MD, Editor

This article will outline two common causes of abdominal pain in children younger than 2 years of age: intussusception and malrotation midgut volvulus. Intussusception refers to invagination or telescoping of a part of the intestine into itself, and malrotation volvulus is a congenital anomaly that results when the normal sequence of rotation and fixation of the bowel fails, which may result in intestinal obstruction.^{6,7} Identifying the cause of bowel obstruction may be as important as recognizing the obstruction itself, as delays in treatment significantly increase morbidity and mortality.^{4,8,9} Both can be challenging to identify and require astute diagnosis and prompt treatment since, ultimately, intestinal obstruction can lead to bowel ischemia, necrosis, perforation, sepsis, shock, and even death.

INTUSSUSCEPTION

Definition

Intussusception refers to invagination or telescoping of a part of the intestine into itself (see Figure 1).⁶ It is a challenging diagnosis and its presentation varies from a well-appearing child with abdominal pain to one who is ill-appearing with altered mental status.

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EXECUTIVE SUMMARY

- Typically, children will complain of severe abdominal pain that is intermittent.
- Intussusception and incarcerated hernia have been found to be the most common causes of acute non-traumatic abdominal pain in children younger than 1 year of age.
- Several recent studies suggest patients can be safely discharged with reliable outpatient follow-up following non-invasive radiologic reduction.
- Morbidity and mortality largely depend on the extent of bowel ischemia, which has been largely attributed to a delay in diagnosis of the malrotation with volvulus.
- Midgut volvulus should be highly considered in any neonate presenting with bilious emesis.
- Upper gastrointestinal (UGI) contrast study to assess the third and fourth part of the duodenum is the gold standard to make the diagnosis; a classic “corkscrew” appearance in the volvulus can be identified with sensitivity of 96%

Epidemiology

Approximately 90% of the time, the diagnosis is made in children younger than 2 years of age and it is fairly uncommon in children less than 3 months.^{1,3} Intussusception is the most common cause of bowel obstruction in infants between 6-36 months of age.¹⁰⁻¹³ Most of the time, the child is healthy with no underlying medical problems, and intussusception tends to affect males slightly more often than females (3:2).¹³

Pathophysiology

The exact etiology is unclear, and 75-90% of the time it is deemed idiopathic.^{4,14} An infectious etiology has been investigated with conflicting results; a virus or bacteria may stimulate lymphatic tissue, resulting in hypertrophy of a Peyer's patch, which can act as a lead point for invagination of the bowel. Theoretically, any pathology within the gastrointestinal (GI) tract may act as a lead point; Meckel's diverticulum and polyps are common culprits.¹⁵⁻¹⁸ There is seasonal predilection associated with gastroenteritis and viral syndromes like upper respiratory tract infection, otitis media, and influenza.¹⁹ Patients with Henoch-Schönlein purpura have a higher incidence of intussusception, which has been proposed to be secondary to areas of intestinal wall hematoma that act as lead points for intussusception. Vaccines have also been postulated as a cause, which led to a previous form of rotavirus vaccine being taken off the market in 1999.²⁰ Two relatively newer studies in 2014 suggest there remains an increase in the rate of occurrence after vaccination.^{20,21}

Other causes in older children are:

Figure 1. Diagram of Intussusception

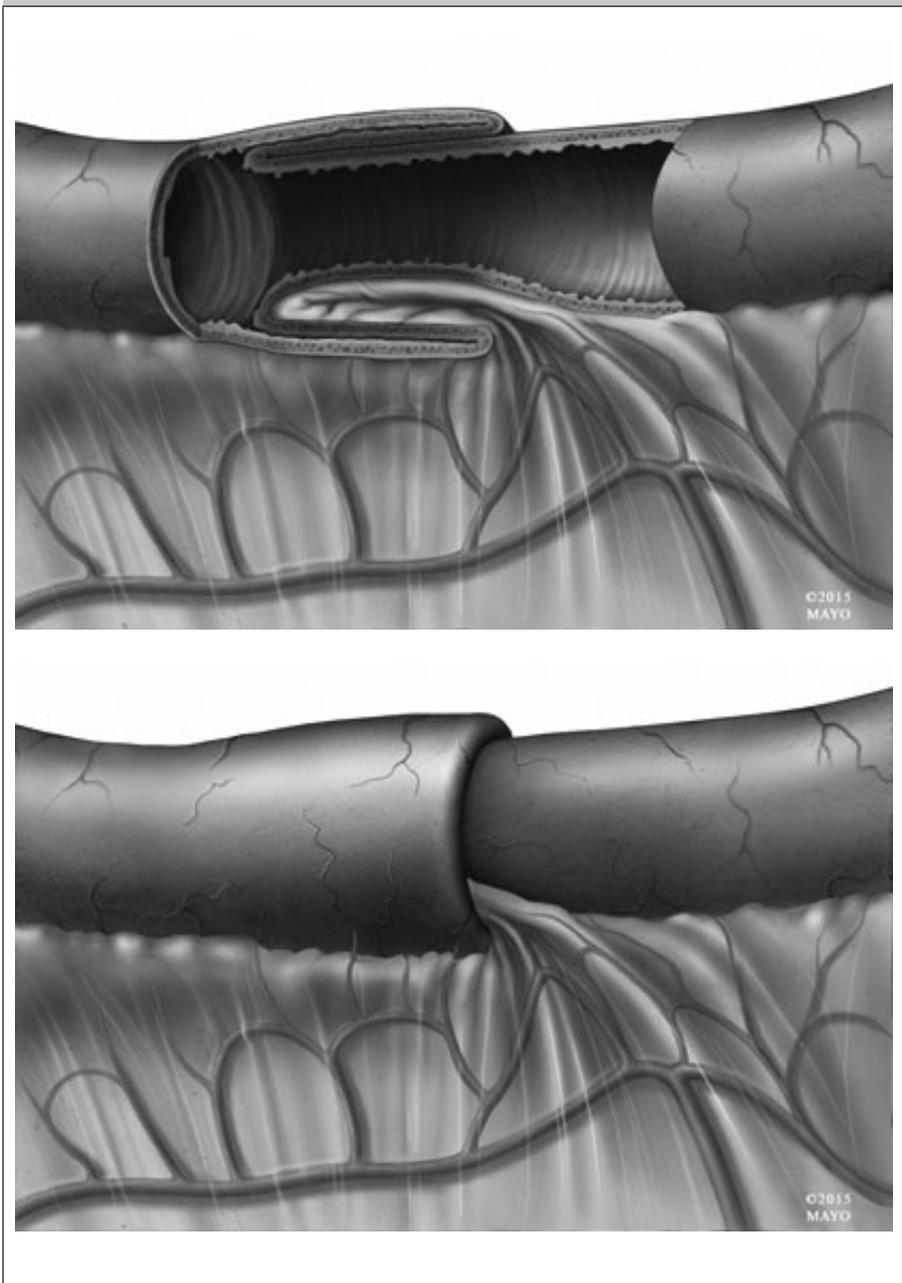


Table 1. Differential Diagnosis for Pediatric Patients Presenting with Altered Mental Status

Altered Mental Status	
A	alcohol acid/base metabolic
E	encephalopathy endocrinopathy
I	insulin (hypoglycemia) intussusception
O	opiates
U	uremia
T	trauma tumor thermal
I	infection intracranial pathology
P	psychogenic poisoning
S	seizure

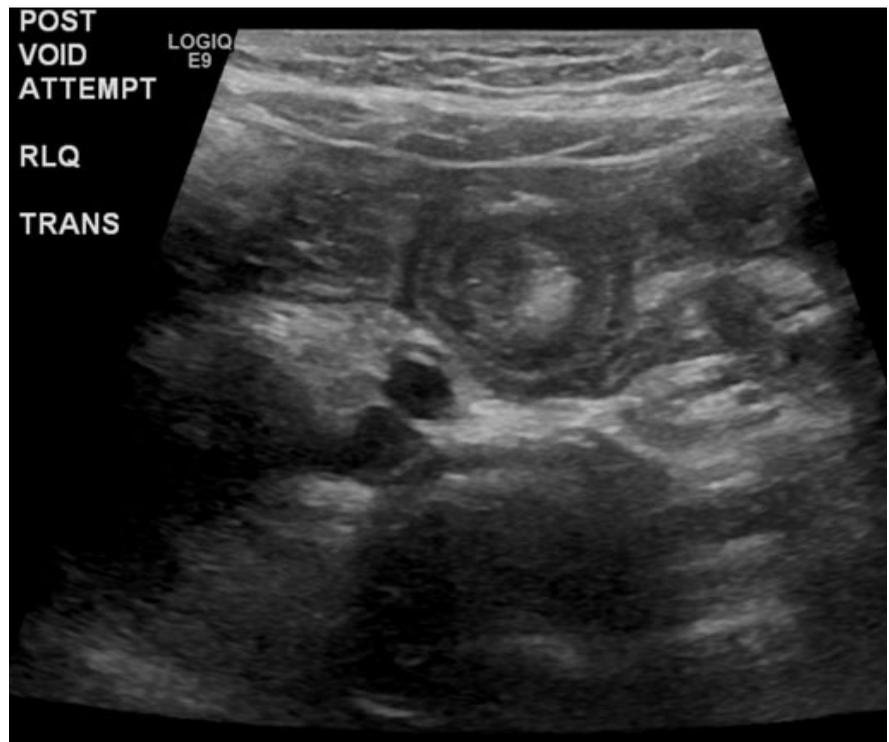
lymphoma, Peutz-Jeghers polyps, inversion of Meckel's, hemangioma, carcinoid, juvenile polyposis coli, and ascaris lumbricoides, among others.

Intussusception occurs when the proximal part of the bowel telescopes into the more distal segment. This invagination can be between virtually any parts of the bowel; however, it occurs most frequently between the ileum and the colon (ileo-colic).^{1,14} The more proximal segment drags portions of the mesentery with it, which contains venous and lymphatic structures leading to congestion, swelling, inflammation, ischemia, necrosis, and eventually perforation.⁵

Clinical Features

Intussusception can be a challenging diagnosis depending on the age of the child and the clinical presentation. Board review books classically depict a young child with colicky abdominal pain, and parents will report currant jelly/red stools, although this is not most commonly seen.¹⁰ Typically, children will complain of severe abdominal pain that is intermittent. As the bowel peristalses and obstructs, children

Figure 2. Ultrasound Depicting Intussusception and "Target Sign"



Source: Mayo Clinic

will experience severe abdominal pain followed by complete resolution of symptoms as the peristaltic obstruction is relieved.²² Later, if the obstruction persists, the bowel can become ischemic and children can become lethargic with an altered mental status. In fact, the differential diagnosis for an altered mental status in children younger than 5 years of age should include intussusception (*see Table 1*).^{13,23}

Children can develop bilious emesis due to the obstruction, and up to 80% might have a mass in the right upper quadrant at the time of presentation. Some children might have fever.²⁴ In the setting of perforation, the abdomen may be distended, rigid, and guarding, and/or rebound tenderness may be elicited.

Diagnostic Studies

The diagnostic approach depends on how the child presents, which is influenced by the patient's age and length of obstruction. More ambiguous cases in the setting of a very ill-appearing

infant will certainly require broad testing, including labs and imaging such as ultrasound and a CT scan of the head and abdomen.²⁵ However, when one suspects intussusception, labs are rarely useful in making the diagnosis.^{12,13} Ultrasound imaging is the gold standard diagnostic modality, and is nearly 100% sensitive and approximately 88-100% specific.^{13,26} A "target sign" can classically be seen on ultrasound as shown in Figure 2. Several studies suggest abdominal X-ray should not be used as a primary screening modality, and emerging data suggest emergency providers working in resource-limited settings may be able to make an accurate diagnosis with bedside ultrasound.^{4,16,24,26-28} Making the diagnosis at the bedside has potential to decrease ED length of stay, expediting management and leading hopefully to better outcomes. However, this is certainly speculative at this time. CT may be considered when ultrasound cannot delineate a true etiology, but it should not be

performed routinely in patients with suspected intussusception.

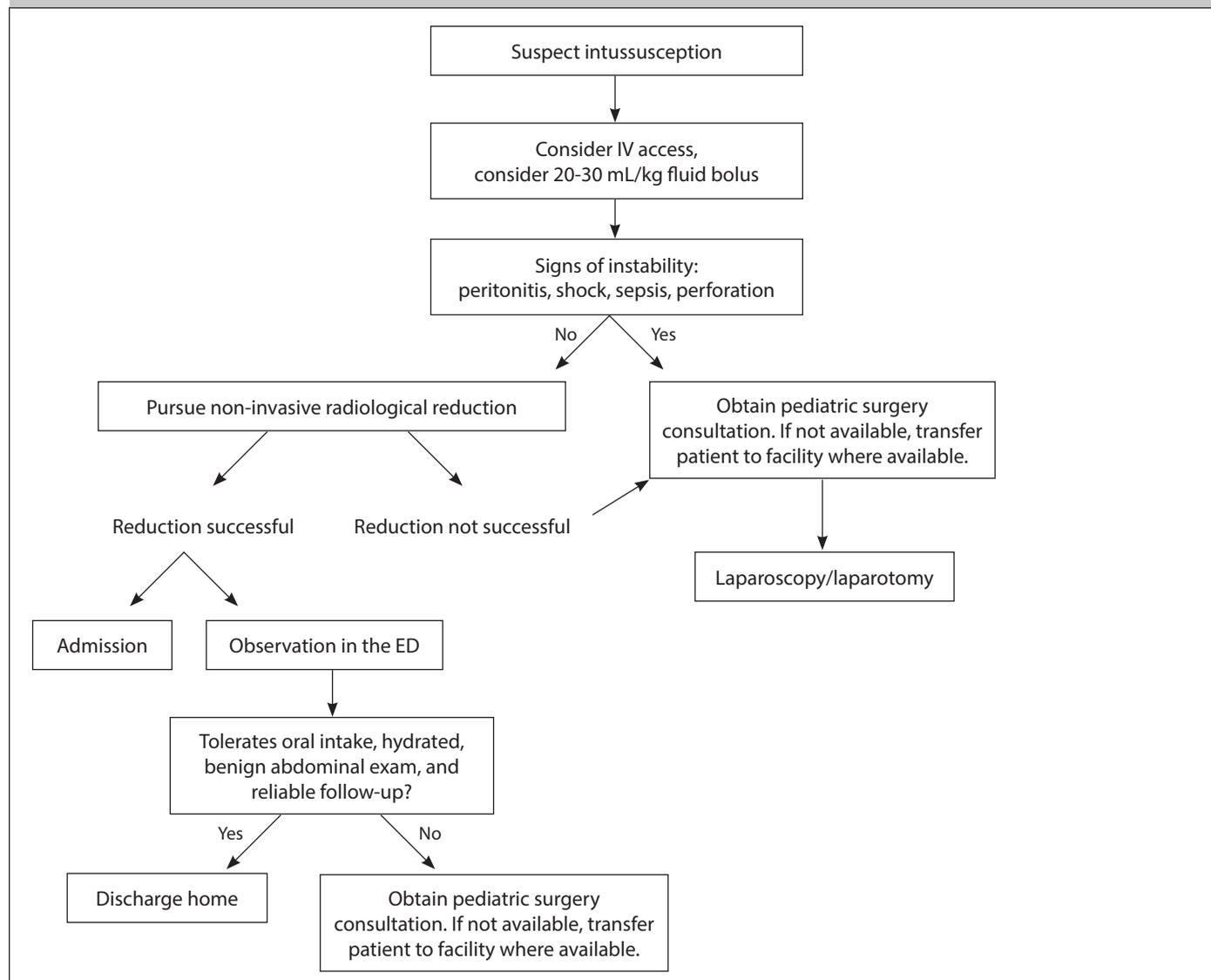
Differential Diagnosis

The differential diagnosis for children with abdominal pain will vary depending on age and clinical presentation (*see Table 2*). Intussusception and incarcerated hernia have been found to be the most common causes of acute non-traumatic abdominal pain in children younger than 1 year of age.¹ However, for any child presenting with altered mental status and abdominal pain, ED physicians must consider intussusception (*see Table 1*). Appendicitis is more commonly found in children older than 1 year of age. Also, one should consider testicular torsion in male patients

Table 2. Differential Diagnosis of Acute Abdominal Pain by Age^{4,5}

< 2 years of age	2-5 years
<ul style="list-style-type: none"> • Colic • Gastroenteritis • Constipation • Urinary tract infection • Intussusception • Volvulus • Incarcerated hernia • Hirschsprung's disease 	<ul style="list-style-type: none"> • Gastroenteritis • Appendicitis • Constipation • Urinary tract infection • Intussusception • Volvulus • Trauma • Incarcerated hernia • Mesenteric adenitis • Testicular torsion • Pharyngitis • Sickle cell crisis • Henoch-Schönlein purpura

Figure 3. Intussusception Management Algorithm



with unexplained abdominal pain and inspection of the genitalia should always be performed.

Management

Up to 90% are fixed and do not reduce spontaneously, requiring reduction (*see Figure 3*). Intravenous access should be considered early, as hypovolemia is common and many will require aggressive fluid resuscitation (20–30 mL/kg isotonic fluid).²⁹ Once the diagnosis is made, a non-invasive radiological reduction should be considered, usually using fluoroscopic guidance with air or contrast. Emerging data suggest radiation-free sonography-guided hydrostatic reduction has a good success rate, although presently it is not standard practice.^{28,30,31} Peritonitis, shock, sepsis, or radiologic evidence of perforation is a contraindication to non-invasive reduction.⁵ The American College of Radiology currently recommends that non-invasive reduction only be performed with a surgeon readily available onsite; however, there have been no large studies validating this recommendation.^{32,33} This information is important for ED providers because following this guideline transfer should be considered promptly once the diagnosis is made if surgical resources are not closely available. A study by Nguyen et al at a large tertiary care children's hospital suggests presence of onsite surgeons may not be necessary, as complications after non-invasive reduction are rare. However, the provider should be comfortable performing percutaneous needle decompression and managing hemodynamic instability, and be able to arrange surgical care if needed. Surgery is performed if non-invasive radiologic reduction is unsuccessful or contraindicated. Reduction can usually be performed laparoscopically, with open laparotomy uncommonly required.

Disposition

Admission is indicated if the patient requires surgical intervention, is critically ill, or has contraindications for non-invasive reduction. However, after successful radiologic reduction, disposition becomes slightly less apparent or universal. Recurrence rates are

approximately 10%, and recurrence itself is not associated with a significant increase in morbidity or mortality.^{34,35} Several recent studies suggest patients can be safely discharged with reliable outpatient follow-up following non-invasive radiologic reduction.^{36–38} For adequately hydrated patients with easily completed non-invasive reduction, observation in the ED for pain recurrence and oral intake is appropriate. If the patient is pain free and can tolerate oral intake, he or she could be discharged home with reliable caregivers. Follow-up and return instructions should be clearly outlined.

If the patient continues to have pain, does not appear adequately hydrated, or if radiologic reduction is difficult, consultation to a pediatric surgeon should be pursued and admission considered.

MALROTATION MIDGUT VOLVULUS

Definition

Intestinal malrotation is a congenital anomaly that results when the normal sequence of rotation and fixation of the bowel fails.^{7,39} Midgut volvulus is a complication of malrotation when the bowel twists around a fixed point usually mesentery that has adhered to the bowel causing obstruction (*see Figure 4*).^{8,40}

Epidemiology

Malrotation is uncommon, and its prevalence in children younger than 1 year of age is 3.9/10,000 live births; however, it can lead to irreversible intestinal necrosis, which can be fatal, and so one must have a high index of suspicion to make the diagnosis.^{13,41,42} Most children present within the first month of age and the majority within the first year of life.³⁹ Morbidity and mortality largely depend on the extent of bowel ischemia, which has been largely attributed to a delay in diagnosis.^{7,9} Males are slightly more affected than females (2:1).^{3,43}

Pathophysiology

During normal embryologic development a counterclockwise turn of both the proximal and midgut

portions of the intestine occurs around the fourth to tenth week of gestation, prior to the intestine retracting into the abdomen.¹³ Abnormalities of rotation physiology result in excessive mobility and compression of the bowel, leading to twisting or volvulus. Mesentery can stick to the bowel and act as a fixed point that the intestine can rotate around, or the bowel may not be fixated correctly to the posterior abdominal wall, which allows the whole midgut to lie free within the abdomen.³⁹ Any part of the intestine may twist and become obstructed, resulting in midgut, cecal, or duodenal volvulus.

Clinical Features

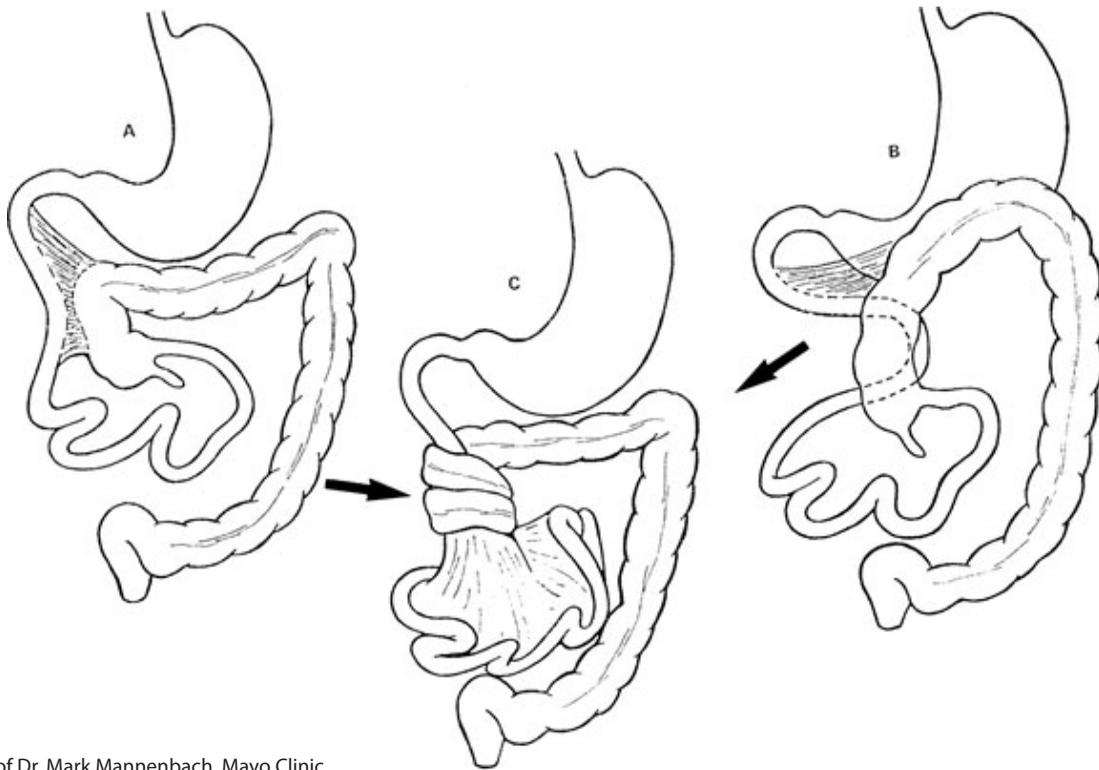
Classically, malrotation with obstruction presents with bilious vomiting, but overall presentation varies.⁴³ Bilious emesis indicates obstruction below the ampulla of Vater, which is a common place for obstruction to occur in the setting of malrotation; although bilious emesis is not pathognomonic for volvulus, it is important to emphasize midgut volvulus should be highly considered in the neonate presenting with bilious emesis.⁹ The age of the infant affects appearance of disease. Neonates may be fussy and parents may complain of feeding difficulties, intermittent apnea, or even failure to thrive. Older infants may appear to have abdominal pain and diarrhea with or without hematochezia and vomiting. Physical exam findings are non-specific; peritoneal signs indicative of perforation that can lead to sepsis and shock are late signs and indicate a poor prognosis.

Diagnostic Studies

The diagnostic studies performed largely depend on the clinical appearance of the child. A broad net with respect to testing is needed in the toxic-appearing infant. Upper gastrointestinal (UGI) contrast study to assess the third and fourth part of the duodenum is the gold standard to make the diagnosis.³⁹ A classic “corkscrew” appearance in the volvulus can be identified with sensitivity of 96%.⁴⁴

Figure 5 shows the X-ray of a newborn presenting with bilious vomiting.

Figure 4. Malrotation Midgut Volvulus



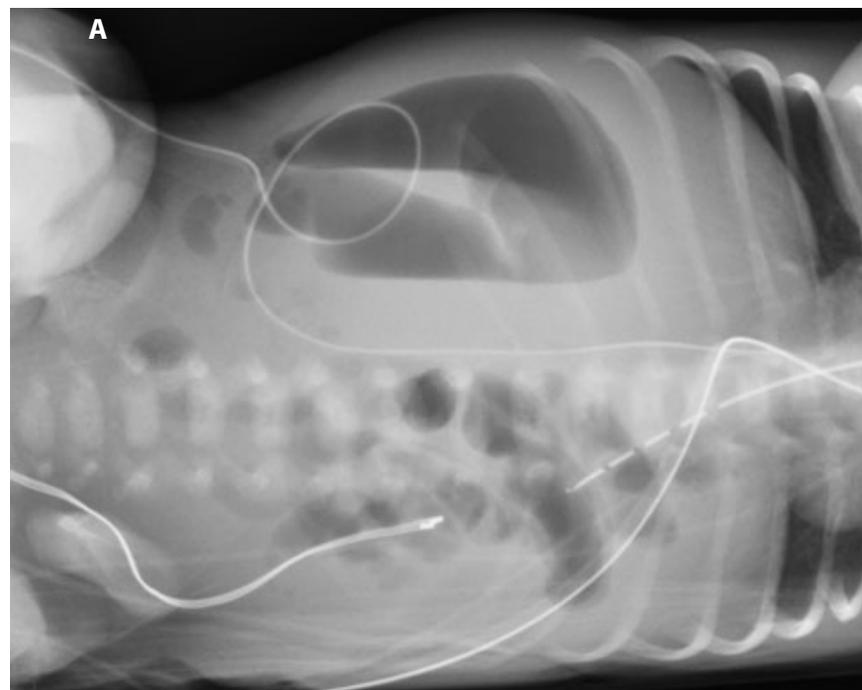
Source: Courtesy of Dr. Mark Mannenbach, Mayo Clinic

The flat and decubitus X-ray shows asymmetry of the bowel gas with distended loops of bowel in the right side and left side of the abdomen. This patient had malrotation with midgut volvulus.

Figure 6 is an upper gastrointestinal study depicting midgut volvulus in a newborn; the study shows that the duodenal jejunal junction crossed to the left of midline but did not extend superiorly as expected, and there was no peristalsis identified within the stomach or duodenum.

Ultrasound of the mesenteric vessels can also be obtained looking for a “whirlpool sign,” which is a swirling shape seen when the superior mesenteric vein (SMV) and mesentery encompass the superior mesenteric artery (SMA),⁴⁵ or for “reversal sign.” Figure 7 is an ultrasound showing SMV/SMA reversal sign in midgut volvulus. The sensitivity and specificity of ultrasound are slightly lower than UGI contrast studies, and, as such, ultrasound usually is used as an adjunct.⁴⁶

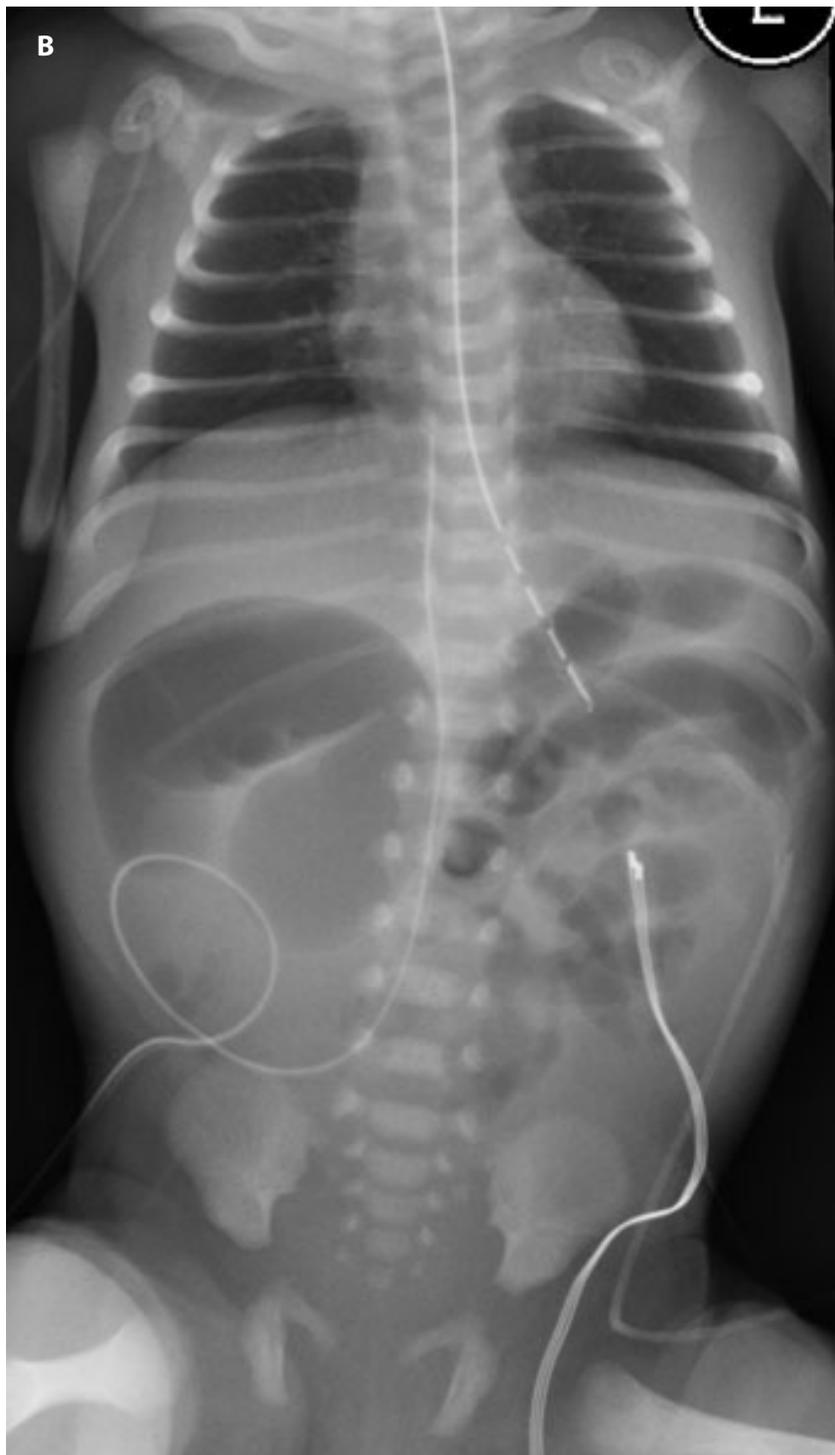
Figure 5. Decubitus (A) of Neonate with Midgut Volvulus



Asymmetry of the bowel gas with a moderately distended loop of bowel in the right and left side of the abdomen. This newborn has midgut volvulus. A Decubitus view

Source: Courtesy of Dr. Mark Mannenbach, Mayo Clinic

Figure 5. X-ray flat (B) of Neonate with Midgut Volvulus



Asymmetry of the bowel gas with a moderately distended loop of bowel in the right and left side of the abdomen. This newborn has midgut volvulus. B Flat view

Source: Courtesy of Dr. Mark Mannenbach, Mayo Clinic

Unstable neonates with sepsis, severe metabolic acidosis, or systemic shock presenting with bilious emesis and abdominal distension should likely forgo imaging and proceed with

surgical exploration. CT scanning is not routinely performed unless the child's presentation is ambiguous or an alternative pathology, such as intra-abdominal mass, is high in the differential.

Differential Diagnosis

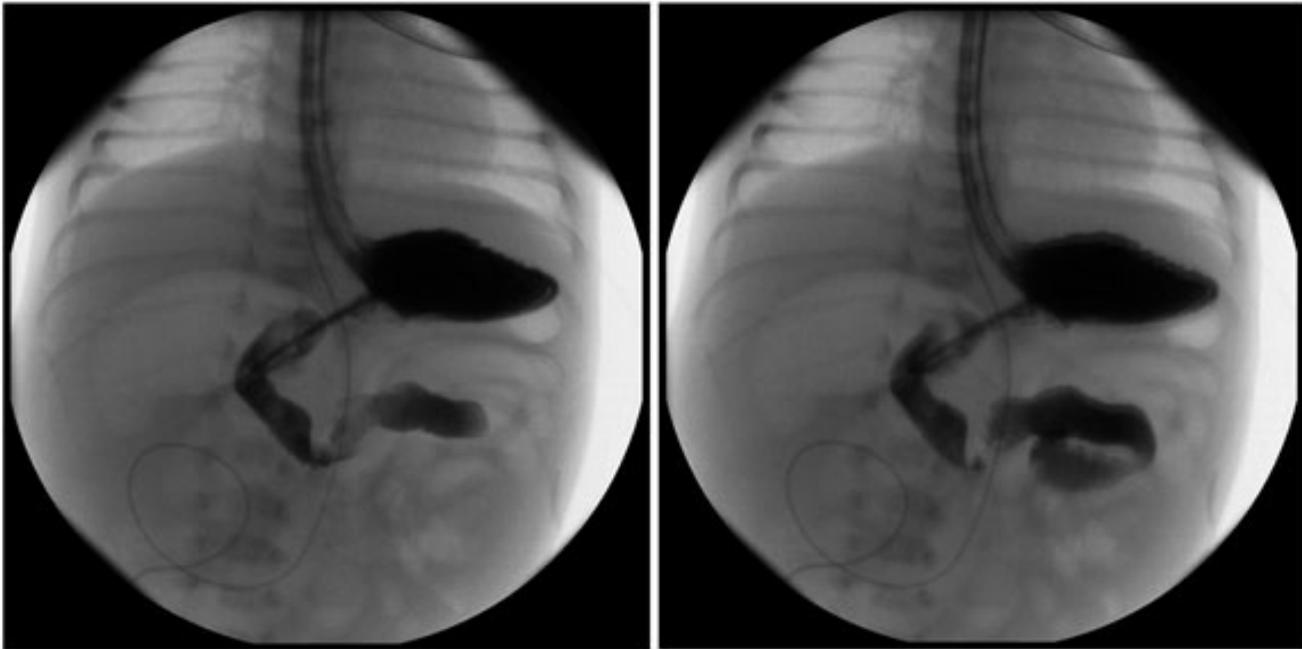
The differential diagnosis for malrotation largely depends on the age of the child, although many will present with abdominal pain (see Table 2.) Malrotation volvulus usually presents in children younger than 1 month of age, so the differential diagnosis will include illnesses more common in this age group. However, one should suspect malrotation midgut volvulus in a child of any age presenting with bilious vomiting and abdominal pain.

Necrotizing enterocolitis should be considered if the child is premature and presents with changes in feeding and abdominal distention. Older infants presenting with signs of intestinal obstruction and altered mental status should raise concern for intussusception.⁴ Pyloric stenosis can present with vomiting; however, vomiting is always non-bilious. It is important to note that malrotation volvulus can present in children of any age and has been diagnosed in adults as well.⁴⁷

Management

Malrotation volvulus is a surgical emergency and requires a laparotomy. Intravenous access should be obtained promptly and aggressive fluid resuscitation started. Nasogastric tube should be placed and antibiotics initiated to cover gram-positive, gram-negative, and anaerobes. Immediate surgical consultation, perhaps even prior to obtaining imaging, may be indicated if the patient is unstable and malrotation volvulus suspected. Morbidity and mortality increase if the obstruction is not treated within 24 hours.³ Surgery is performed to correct the obstruction and minimize risk of future volvulus; the surgery will not correct the actual malpositioning of the bowel. The surgical approach, laparotomy or laparoscopic, does not affect length of stay or the complication rate.⁴⁸ If bowel necrosis is found, bowel resection will occur, placing the patient at risk for short bowel syndrome, again, emphasizing the need for prompt diagnosis and treatment. The longer the bowel stays obstructed the more likely ischemia and necrosis ensue.

Figure 6. Upper Gastrointestinal Study Depicting Midgut Volvulus in a Newborn



The duodenal jejunal junction crossed to the left of midline but did not extend superiorly as expected, and there was no peristalsis identified within the stomach or duodenum.

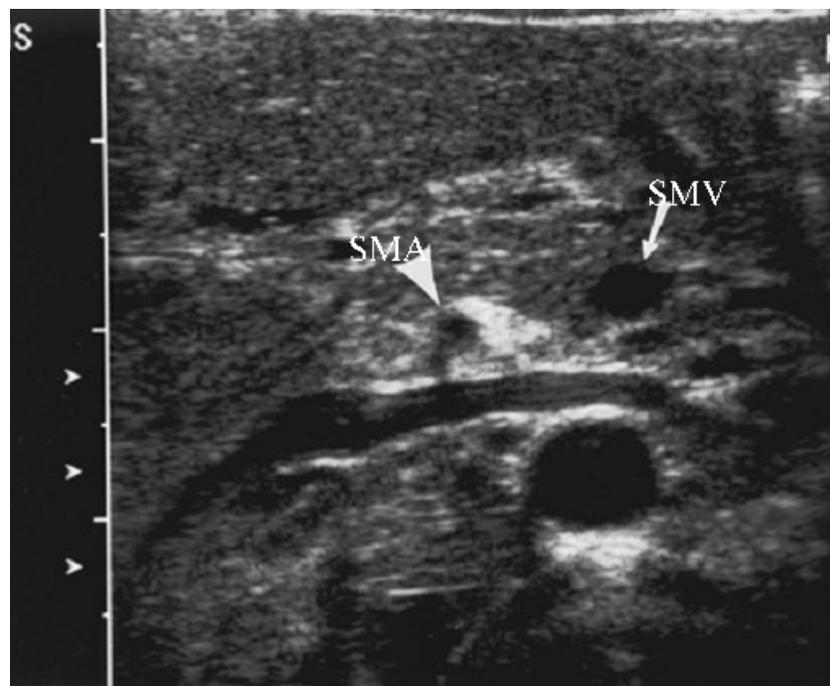
Disposition

Children diagnosed with malrotation volvulus require admission at a facility with pediatric surgical resources. Transfer should be considered even prior to diagnosis if malrotation volvulus is suspected.

Summary

Intussusception is usually diagnosed in children younger than 2 years of age. It affects males slightly more often than females. The exact etiology is unclear; however, there is an association with gastroenteritis and viral syndromes such as upper respiratory tract infection, otitis media, and influenza. Most frequently, the intussusception occurs between the ileum and the colon. Children can present with intermittent abdominal pain, vomiting, and currant jelly or red stools; however, the latter is a late sign. Intussusception should be considered in a child presenting with altered mental status. Non-invasive radiological reduction should be attempted; however, if the child appears critically ill, pediatric surgery should be consulted promptly.

Figure 7. Ultrasound Showing SMV/SMA Reversal Sign in Midgut Volvulus



Source: Courtesy of Dr. Mark Mannenbach, Mayo Clinic

Malrotation midgut volvulus most commonly presents in children younger than 1 month of age. Bilious

emesis in a neonate should be considered a surgical emergency until proven otherwise. Patients can present

with irritability, feeding difficulties, failure to thrive, and abdominal pain. The diagnosis can be challenging and delays in diagnosis increase morbidity and mortality significantly. Signs of bowel ischemia, such as hematochezia or sepsis, indicate a poor prognosis. UGI contrast study is the gold standard diagnostic modality, and pediatric surgical consultation is required since an operation is required.

Overall, children presenting to the ED with abdominal pain can be challenging to diagnose. ED physicians are charged to differentiate between self-limited pathology and more life-threatening surgical emergencies. Intussusception and malrotation mid-gut volvulus both can present with abdominal pain, vomiting, and irritability. Both can be fatal if the diagnosis is delayed or missed, and so one must have a high index of suspicion.

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CME Questions

1. What is the most common age range for intussusception?
 - a. Children younger than 2 years of age
 - b. Children age 5 to 8 years of age
 - c. Children age 10 to 15 years of age
 - d. None of the above
2. Which of the following is true regarding intussusception?
 - a. It has been associated with a viral illness.
 - b. It has been associated with rotavirus vaccination.
 - c. It is most frequently located in the ileocolic region.
 - d. All are of the above
3. Which of the following is characteristic of a child with intussusception?
 - a. Colicky abdominal pain
 - b. Currant jelly/red stools
 - c. Fever
 - d. Both a and b
 - e. All of the above
4. The gold standard for the diagnosis of intussusception is:
 - a. ultrasound.
 - b. complete blood count showing left shift.
 - c. computed tomography.
 - d. X-ray of the abdomen.
5. The management of intussusception includes:
 - a. IV for analgesia and fluid resuscitation.
 - b. non-invasive radiologic intervention using fluoroscopic guidance with air or contrast enema.
 - c. surgery if non-invasive radiologic reduction is unsuccessful or contraindicated.
 - d. All of the above
6. Contraindications for enema reduction include which of the following?
 - a. Peritonitis
 - b. Shock
 - c. Sepsis
 - d. Suspected perforation
 - e. All of the above
7. Which of the following is true regarding malrotation volvulus?
 - a. It is uncommon.
 - b. It can be fatal.
 - c. Most children present within the first month of life.
 - d. All of the above
8. Which of the following are characteristic of a neonate with malrotation volvulus?
 - a. Bilious vomiting
 - b. Feeding difficulties
 - c. Failure to thrive
 - d. Apnea
 - e. All of the above
9. The management of malrotation volvulus includes:
 - a. emergent surgery.
 - b. elective surgery.
 - c. fluoroscopy guided air enema.
 - d. ultrasound of the bowel to rule out obstruction.
10. Which of the following is true regarding intussusception and malrotation volvulus?
 - a. The chief complaint can be "fussiness."
 - b. Children can be critically ill and lethargic.
 - c. Patients have abdominal pain or distension.
 - d. All of the above

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CME Objectives

Upon completion of this educational activity, participants should be able to:

- recognize specific conditions in pediatric patients presenting to the emergency department;
- describe the epidemiology, etiology, pathophysiology, historical and examination findings associated with conditions in pediatric patients presenting to the emergency department;
- formulate a differential diagnosis and perform necessary diagnostic tests;
- apply up-to-date therapeutic techniques to address conditions discussed in the publication;
- discuss any discharge or follow-up instructions with patients.

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