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Neonatal Intestinal Obstruction

Intestinal obstructions are the most common surgical emergencies in the neonatal period. Early and accurate diagnosis of intestinal obstruction is paramount for proper patient management. For evaluation and diagnosis, intestinal obstruction in neonates can be divided into either high or low obstruction on the basis of the number of dilated bowel loops present on the initial abdominal radiographs. Although three or fewer dilated bowel loops are typically seen with high intestinal obstruction, more than three are generally seen with low intestinal obstruction in neonates. High intestinal obstructions are defined as occurring proximal to the ileum, resulting in various combinations of gastric, duodenal, and jejunal dilatation according to the level of obstruction (Table 1). In contrast, low intestinal obstructions involve the distal ileum or colon and typically result in diffuse dilatation of multiple small-bowel loops (Table 1). Although neonates with classic radiographic findings of high intestinal obstruction, such as duodenal atresia, may directly undergo surgery without any additional imaging, an upper gastrointestinal series is typically performed for further evaluation. Similarly, an enema examination is used for further investigation of low intestinal obstruction in neonates.

High Intestinal Obstruction

Gastric Atresia

Complete gastric agenesis virtually never occurs. A congenitally small stomach, or microgastria, is rare. It can present as an isolated malformation or with other anomalies, especially with heterotaxia syndrome (i.e., abnormal left-right asymmetry) with asplenia. The characteristic imaging findings of microgastria include a distended esophagus and a small midline stomach. Gastric atresia, which is typically localized to the antrum or pyloric region, is also rare, accounting for < 1% of all congenital intestinal obstructions. The cause of gastric atresia is presumably related to a localized intrauterine vascular occlusion rather than developmental failure of bowel canalization. Affected newborns usually present with nonbilious vomiting and abdominal distention within the first few hours after birth. Abdominal radiographs in neonates with distal gastric atresia are characterized by a gas-filled markedly dilated stomach without distal intestinal air. This imaging finding is known as the "single bubble sign."

Duodenal Atresia

Complete duodenal obstruction in neonates is usually caused by duodenal atresia, which results from congenital failure

TABLE 1: Causes of Neonatal Intestinal Obstruction

High intestinal obstruction
Gastric atresia
Duodenal atresia
Duodenal stenosis (with annular pancreas)
Duodenal web
Malrotation
Jejunal atresia and stenosis
Low intestinal obstruction
Small bowel involvement
Ileal atresia
Meconium ileus
Large bowel involvement
Functional immaturity of the colon
Hirschsprung disease
Colonic atresia
Anal atresia and anorectal malformations

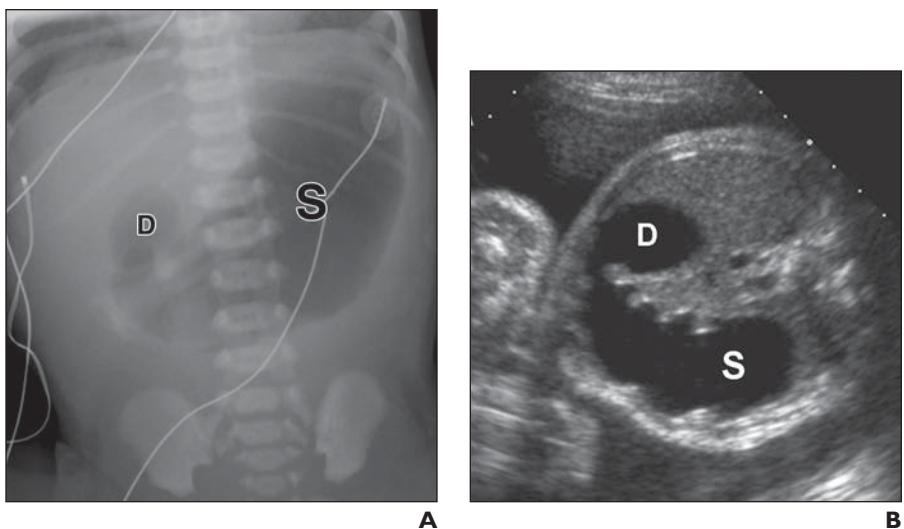


Fig. 1—Duodenal atresia in newborn girl with vomiting and abdominal distention. **A**, Abdominal radiograph shows gas-filled distended stomach (S) and duodenal bulb (D), producing “double bubble sign.” **B**, Prenatal transverse ultrasound image shows fluid-filled distended stomach (S) and duodenal bulb (D), producing prenatal ultrasound double bubble sign.

of recanalization that normally occurs during 9–11 weeks of gestational age. In contrast to more distal small-bowel atresias, duodenal atresia does not appear to be related to intrauterine vascular insults. Duodenal atresia is frequently associated with other congenital anomalies, such as additional intestinal atresias, congenital heart disease, or as a part of the VACTERL association (i.e., vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies). Approximately 30% of cases occur in patients with the diagnosis of trisomy 21 (Down syndrome). Neonates with duodenal atresia typically present with vomiting during the first several hours of life. In approximately 80% of affected neonates, the site of duodenal atresia is postampullary, so that the patient may present with bilious emesis. Abdominal radiographic findings of duodenal atresia include a gas-filled dilated stomach and duodenal bulb, known as the classic “double bubble sign” (Fig. 1). In cases of complete duodenal atresia, there is always a lack of bowel gas distal to the proximal duodenum.

Duodenal Stenosis (With Annular Pancreas)

Partial duodenal obstruction in neonates is usually caused by duodenal stenosis, with or without annular pancreas. As in neonates with duodenal atresia, there is gastroduodenal distention, but in duodenal stenosis there is also intestinal gas seen distal to the proximal duodenum. On upper gastrointestinal studies, the duodenal bulb is distended and there is slow transit of oral contrast material through the stenotic segment of the duodenum to the distal bowel (Fig. 2). Because of the partial obstruction, affected pediatric patients may present later in life than those with duodenal atresia (Fig. 3).

Duodenal Web

“Duodenal web” refers to a small congenital obstructing membrane with a central pinhole aperture that constitutes a functional web. Long-term pressure of peristalsis against the stenotic segment of the duodenum may lead to distal stretching of the web, forming an intraluminal pseudodiverticulum (windsock diverticulum). The characteristic upper gastrointestinal finding is a faint radiolucent membrane, which represents with barium filling the lumen and around the diaphragm (Fig. 4).

Malrotation

Malrotation refers to abnormal or incomplete intestinal rotation during embryonic development. Normally, physiologic bowel rotation (270° around the axis of the superior mesenteric artery) leads to duodenal-jejunal and ileocecal junctions that are appropriately situated

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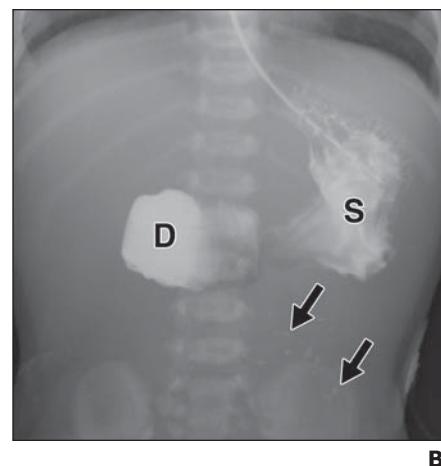
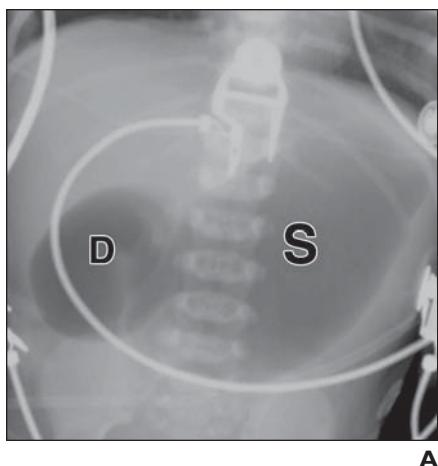


Fig. 2—Duodenal stenosis in 4-day-old girl with vomiting and abdominal distention. **A**, Abdominal radiograph shows severe distention of stomach (S) and duodenal bulb (D). **B**, Delayed abdominal radiograph from upper gastrointestinal study shows barium-filled stomach (S) and distended duodenum (D). Trickles of barium (arrows) have slowly progressed into more distal portions of bowel.

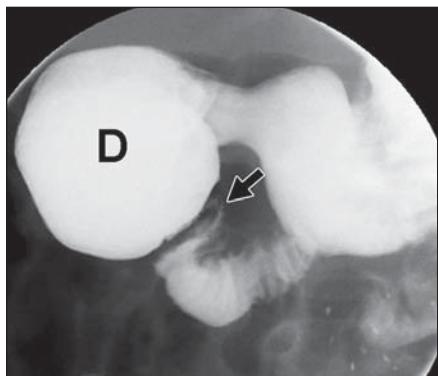


Fig. 3—Duodenal stenosis in 3-month-old girl with progressively worsening vomiting and abdominal distention. Image from upper gastrointestinal study shows severely distended barium-filled duodenal bulb (D). Barium passes through stenotic segment (arrow) of duodenum into more distal portions of bowel.

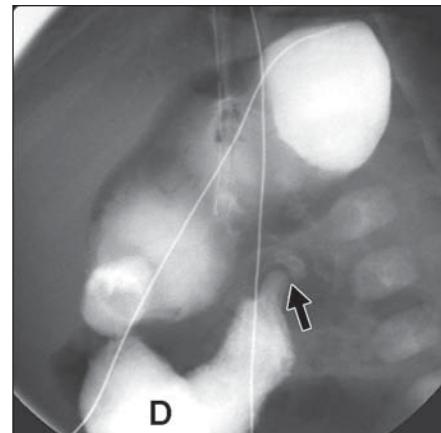


Fig. 4—Duodenal web in 3-day-old boy with vomiting and abdominal distention. Image from upper gastrointestinal study shows faint radiolucent obstructing membrane (arrow) with barium-filled duodenal lumen. D = duodenal bulb.

in the left upper and right lower quadrants, respectively. This normal physiologic bowel rotation results in a broad attachment of the intestines to the mesentery, which prevents bowel from twisting around the mesentery. In malrotation, different degrees of abnormal location of these two key intestinal segments produce a narrower mesenteric attachment, which places the small bowel at risk for twisting around its pedicle, a condition known as “midgut volvulus.” Midgut volvulus causes both mechanical obstruction and arterial occlusion of the mesenteric vessels. If untreated, midgut volvulus will progress to bowel ischemia and eventual infarction. Furthermore, a malpositioned portion of intestine is often associated with abnormal peritoneal fibrous bands (Ladd bands), which are anomalous fibrous connections that typically extend from the malpositioned cecum across the duodenum to attach to the peritoneum and liver. These anomalous fibrous bands may also contribute to bowel obstruction or be solely responsible for it.

Although a newborn with isolated malrotation may be completely asymptomatic, the development of midgut volvulus typically produces bilious emesis. The abdominal radiograph of a neonate with malrotation is commonly nonspecific. It may be entirely normal, show a

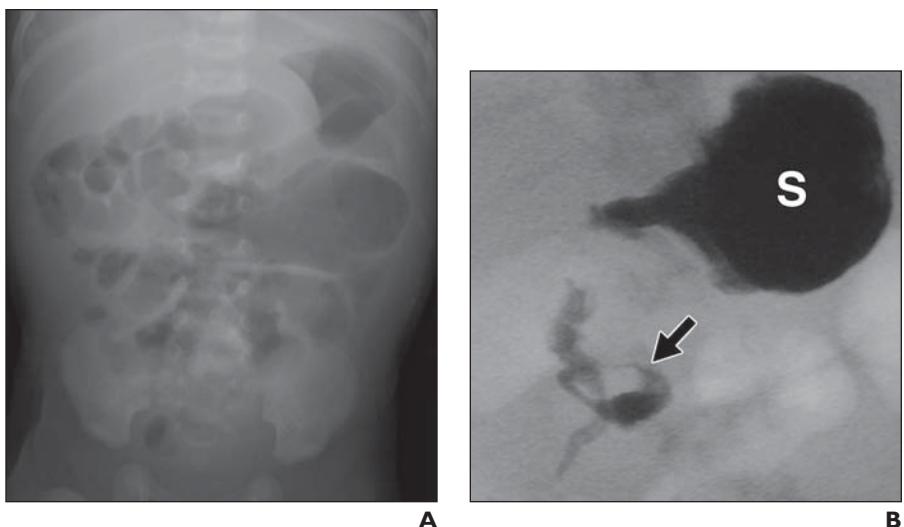


Fig. 5—Malrotation with volvulus in 11-day-old boy with bilious vomiting for 1 day.

A, Abdominal radiograph shows nonspecific dilatation of several bowel loops.

B, Image from upper gastrointestinal study shows abnormal course of duodenum (arrow), which fails to cross midline and has spiral (corkscrew) appearance, consistent with malrotation with volvulus. **S** = stomach.

proximal bowel obstruction pattern, or show dilatation of multiple bowel loops (Fig. 5A). In these instances, further evaluation requires an upper gastrointestinal study as the next imaging procedure. The classic upper gastrointestinal appearance of malrotation with volvulus consists of an abnormal course of the duodenum that fails to cross the midline combined with a circular duodenal configuration (corkscrew appearance) (Fig. 5B).

Jejunal Atresia and Stenosis

Obstruction of the jejunum is virtually always due to atresia or stenosis in neonates. The underlying cause of jejunal and ileal atresias is an intrauterine ischemic insult, which can be a primary vascular cause or secondary to a volvulus. Newborns with jejunal atresia usually present with bilious emesis and abdominal distention. Abdominal radiography typically shows several dilated bowel loops, more than the double bubble of duodenal atresia but fewer than the low obstruction pattern.

Characteristic dilatation of the stomach, duodenum, and proximal jejunum is known as the “triple bubble sign” (Fig. 6A). The loop of jejunum immediately proximal to the atresia is frequently disproportionately dilated and has a bulbous end. In most instances, an upper gastrointestinal study is not necessary before surgical repair. However, if obtained, an upper gastrointestinal study usually shows a dilated duodenum and proximal jejunum, both of which are filled with contrast material. A contrast enema study also is often performed to evaluate for possible concomitant distal bowel atresia before surgical repair (Fig. 6B).

Low Intestinal Obstruction

Small-Bowel Involvement

Ileal atresia—Ileal atresia is a common cause of low intestinal obstruction in neonates, with an estimated incidence of 1 in 5000 live births. The cause is thought to be related to an intrauterine ischemic insult, similar to the more proximal small-bowel atresias. The distal portion of the ileum is most commonly involved. Neonates with ileal atresia have fewer associated congenital anomalies than those with duodenal atresia. Affected patients usually present with bilious vomiting and abdominal distention. As in other causes of low intestinal obstruction in neonates, abdominal radiography usually shows numerous dilated bowel loops (Fig. 7A). A contrast enema study shows termination of the contrast-filled colon near the distal ileum (in the case of distal ileal atresia), associated with multiple air-filled distended small-bowel loops (Fig. 7B).

Meconium ileus—Meconium ileus accounts for approximately 20% of cases of neonatal intestinal obstruction. Caused by intraluminal obstruction of the colon and distal small bowel

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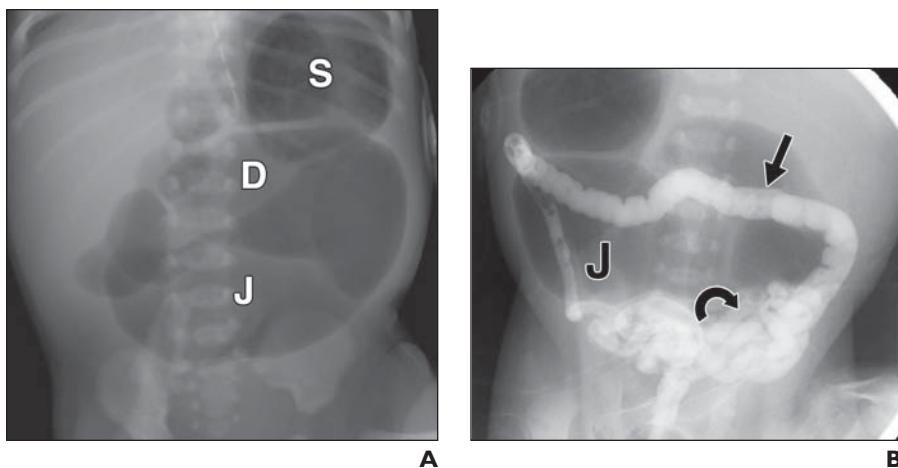


Fig. 6—Jejunal atresia in newborn girl with prenatal diagnosis of intestinal obstruction and vomiting. **A**, Abdominal radiograph shows severe dilatation of stomach (S), duodenum (D), and jejunum (J), producing “triple bubble sign.” Note lack of distal intestinal air. **B**, Image from contrast enema shows microcolon (straight arrow) and ileum (curved arrow), as well as severely dilated jejunum (J).

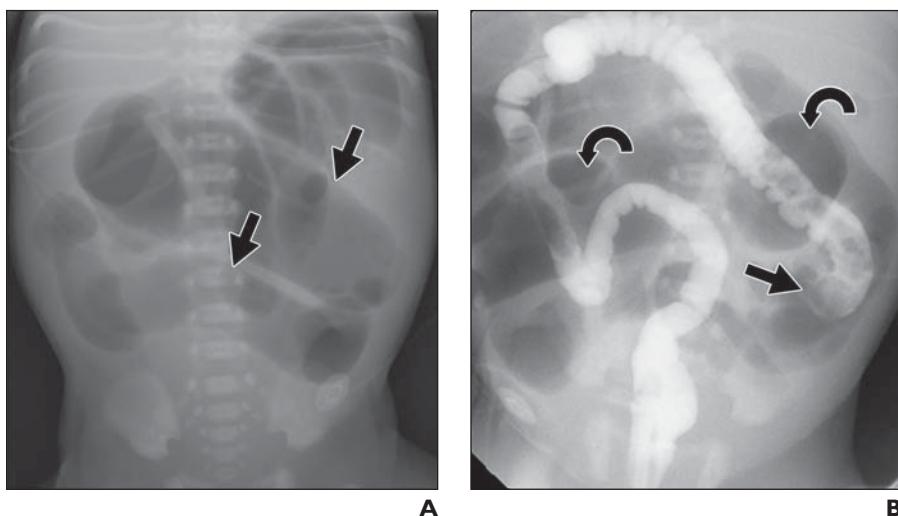


Fig. 7—Ileal atresia in 2-day-old boy with progressively worsening vomiting and abdominal distention. **A**, Abdominal radiograph shows multiple distended bowel loops (arrows). Note lack of distal intestinal air. **B**, Image from contrast enema study shows contrast-filled colon ending near distal ileum (straight arrow). There are multiple distended loops of air-filled small bowel (curved arrows).

from abnormal concretions of meconium, this condition is virtually always the earliest clinical manifestation of cystic fibrosis. Occlusion of the distal small bowel results in mechanical obstruction with subsequent distention of the more proximal bowel loops. Meconium ileus may be complicated by volvulus, perforation, or peritonitis. Intrauterine perforation may lead to meconium peritonitis (chemical type), and peritoneal calcifications may be seen postnatally (Fig. 8). Abdominal radiographs in neonates with meconium ileus usually show a pattern of low intestinal obstruction that is characterized by multiple bowel loop dilatations (Fig. 9A). Despite the marked intestinal dilatations, there is often a relative lack of air-fluid levels within the dilated bowel loops because of the abnormally thick intraluminal meconium. A contrast enema study typically shows an unused colon (i.e., microcolon), within which are multiple small filling defects representing meconium concretions. If there is reflux of contrast material beyond the ileocecal valve, multiple small filling defects (meconium concretions) also may be seen in the terminal ileum (Fig. 9B).

Fig. 8—Meconium peritonitis in 2-day-old boy with intrauterine meconium perforation who presented with enlarged scrotum and rigid abdomen. Abdominal radiograph shows multiple peritoneal calcifications (straight arrows). Note also calcification (curved arrow) in scrotum.

Large-Bowel Involvement

Functional immaturity of the colon (meconium plug or small left colon syndrome)—Functional immaturity of the colon, also known as meconium plug or small left colon syndrome, is a typically benign and self-limited transient functional colonic obstruction in neonates. The underlying cause is thought to be related to immaturity of the colonic ganglion cells (myenteric nerve plexus). Functional immaturity of the colon is the most common diagnosis in neonates who fail to pass meconium stool for more than 48 hours. An increased incidence of functional immaturity of the colon has been reported in infants of diabetic mothers and mothers who received magnesium sulfate for preeclampsia during pregnancy.

Abdominal radiographs in neonates with functional immaturity of the colon typically show multiple dilated bowel loops, an appearance that is characteristic for low intestinal obstruction (Fig. 10A). On a contrast enema study, multiple filling defects (i.e., meconium plugs) may be seen, although the amount of colonic meconium is variable and discrete meconium plugs may not be present. The ascending and transverse portions of the colon are typically more dilated than the descending colon (i.e., small left colon syndrome), whereas the rectum is normal in size (Fig. 10B). A diagnostic enema is usually therapeutic and leads to the passage of discrete meconium plugs and resolution of the intestinal obstruction.

Despite the characteristic contrast enema pattern of functional immaturity of the colon in neonates, an important differential consideration is Hirschsprung disease (see next section). Unusual cases of Hirschsprung disease that involve a long segment and have a transition point

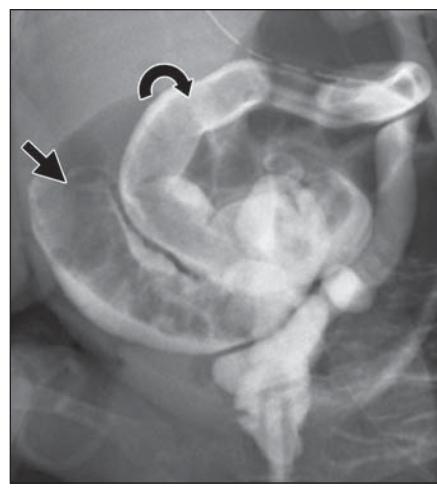
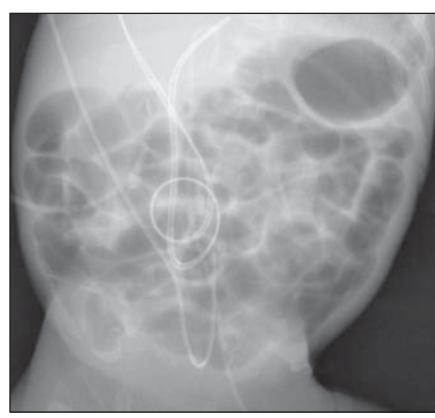


Fig. 9—Meconium ileus in 3-day-old girl who failed to pass meconium.

A, Abdominal radiograph shows multiple dilated bowel loops.

B, Image from contrast enema shows large amount of meconium concretions in markedly distended terminal ileum (straight arrow) and colon (curved arrow).

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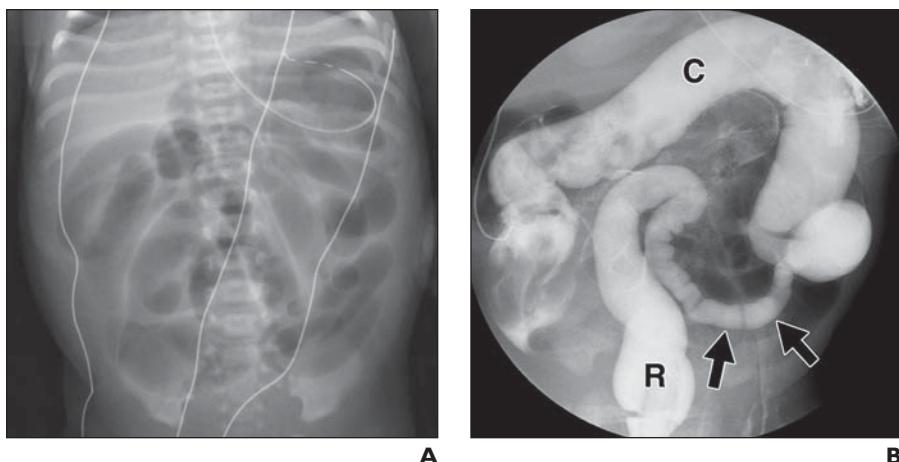


Fig. 10—Functional immaturity of colon in newborn boy who failed to pass meconium and **had** abdominal distention.

A, Abdominal radiograph shows multiple dilated bowel loops.

B, Contrast enema shows that left colon is small (arrows), whereas remaining colon (C) and rectum (R) are normal in size.

at the splenic flexure may completely mimic functional immaturity of the colon on imaging studies. Therefore, neonates with a presumed diagnosis of functional immaturity of the colon whose symptoms do not resolve after therapeutic enema should be considered for colonic biopsy to make a definitive diagnosis.

Hirschsprung disease—Hirschsprung disease is caused by an arrest of neuronal (ganglion) cell migration to the distal bowel before the 12th week of gestational age. Because the intestinal ganglion cells migrate in a craniocaudal direction, the area of aganglionosis always extends distally from the point of neuronal arrest to the anus. The segment of aganglionosis is typically continuous and most commonly involves the rectum and a portion of the sigmoid colon (short segment disease, which accounts for about 75% of cases).

Occasionally, the migration abnormality of ganglion cells is more extensive and the aganglionic segment extends for a variable distance proximal to the sigmoid colon (long segment disease, which accounts for about 25% of cases). The process can also affect the entire colon (total colonic aganglionosis) and, rarely, a portion of the small intestine. The aganglionic segment is unable to distend normally, resulting in a functional obstruction with proximal bowel dilatation and abnormal defecation. Most cases present clinically in the newborn period with delayed passage of meconium and abdominal distention.

Abdominal radiographs in neonates with Hirschsprung disease typically show a pattern of low intestinal obstruction with dilatation of numerous bowel loops (Fig. 11A). This appearance indicates low intestinal obstruction but is not specific. Therefore, a contrast enema study usually is obtained subsequently for further evaluation. Characteristic radiologic findings of Hirschsprung disease on an enema study include an abnormal rectosigmoid ratio (< 1) (Fig. 11B), transition zone of rectal narrowing (Fig. 11B), irregular rectal contractions (Fig. 11C), and retained contrast material on delayed radiographs (Fig. 11D). Although the contrast enema study is typically abnormal in neonates with Hirschsprung disease, it may be completely normal. Therefore, the possibility of Hirschsprung disease still should be strongly suspected, and rectal biopsy is usually required for definitive diagnosis in a neonate with clinical signs and symptoms of persistent low intestinal obstruction but a normal contrast enema.

Colonic atresia—The colon is a relatively uncommon site for intestinal atresia, with an estimated incidence of 5–15% of all intestinal atresias in neonates. It usually results from an intrauterine vascular insult and is typically located in the colon proximal to splenic flexure. Affected neonates generally present with abdominal distention. Abdominal radiographs in neonates with colonic atresia typically show multiple dilated bowel loops, multiple air–fluid levels, and absence of air in the rectum. Occasionally, there may be a disproportionately distended loop that represents the most distal aspect of the colonic atresia, a finding that should suggest the diagnosis (Fig. 12A). A contrast enema study typically shows a distal unused colon (i.e., microcolon), with the more proximal markedly dilated colon ending in a blind pouch (Fig. 12B).

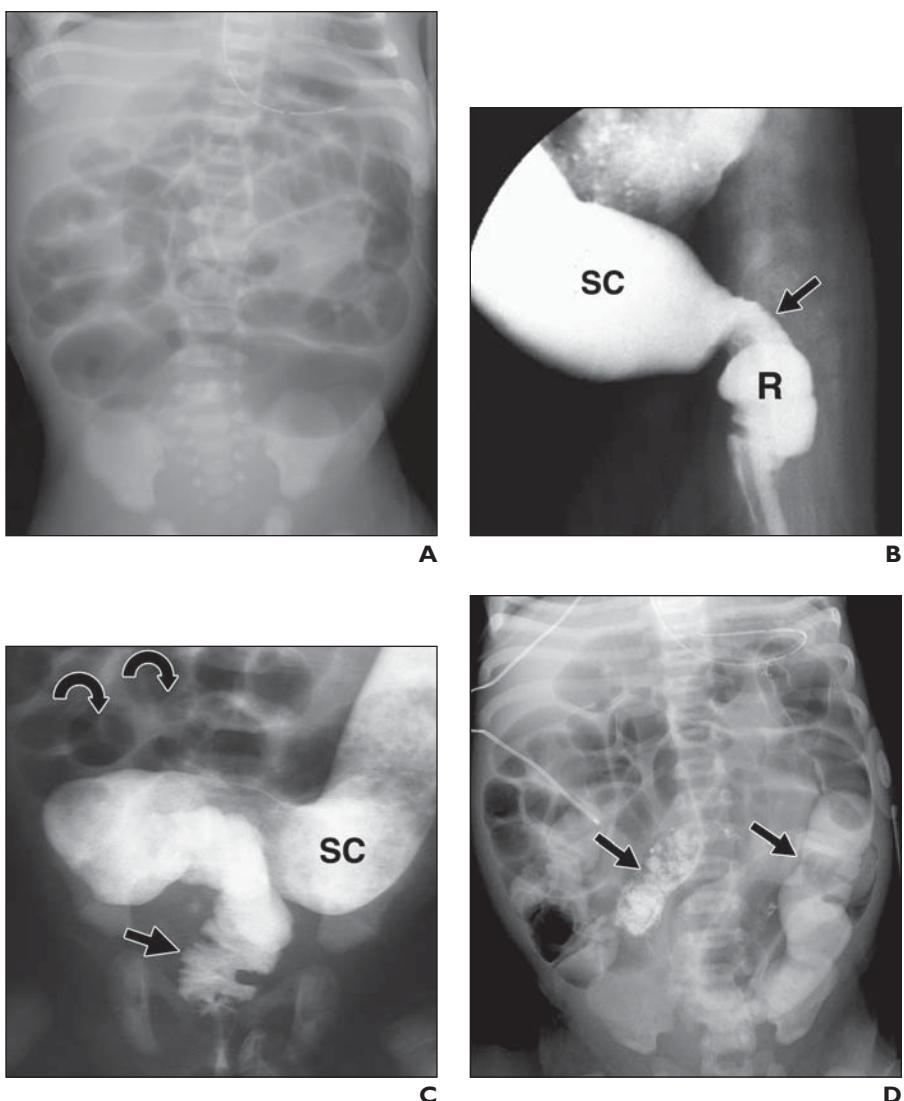


Fig. 11—Hirschsprung disease in 2-day-old boy who failed to pass meconium and had abdominal distention. **A**, Abdominal radiograph shows multiple dilated bowel loops and lack of air in region of rectum. **B**, Image from contrast enema shows area of rectal narrowing (arrow). Note that size of sigmoid colon (SC) is larger than that of rectum (R). **C**, Image from contrast enema study shows irregular rectal contractions (straight arrow). Note multiple distended loops of small bowel (curved arrows). SC = sigmoid colon. **D**, Delayed abdominal radiograph obtained 24 hours after contrast enema study shows retained contrast material within multiple loops of dilated bowel (arrows).

Anal atresia and anorectal malformations—Anal atresia, also known as “imperforate anus,” is a condition of unknown cause in which there is the absence of a normal anal opening. With an estimated incidence of 1 in 5000 live births, anal atresia affects boys and girls with similar frequency. Anal atresia is associated with other congenital anomalies, including vertebral, cardiac, renal, and limb anomalies.

Although there are currently numerous classification schemes for anal atresia, it is traditionally divided into high and low lesions, depending on whether the rectum ends above or below the puborectalis sling. In a low anal atresia with the rectum ending below the puborectalis sling, the rectum remains close to the skin in a blind pouch. The anus may be stenotic or not even present on physical examination. In a high anal atresia with the rectum ending above the puborectalis sling, the rectum is located high in the pelvis and there may be a fistulous connection between it and the bladder, urethra, or vagina.

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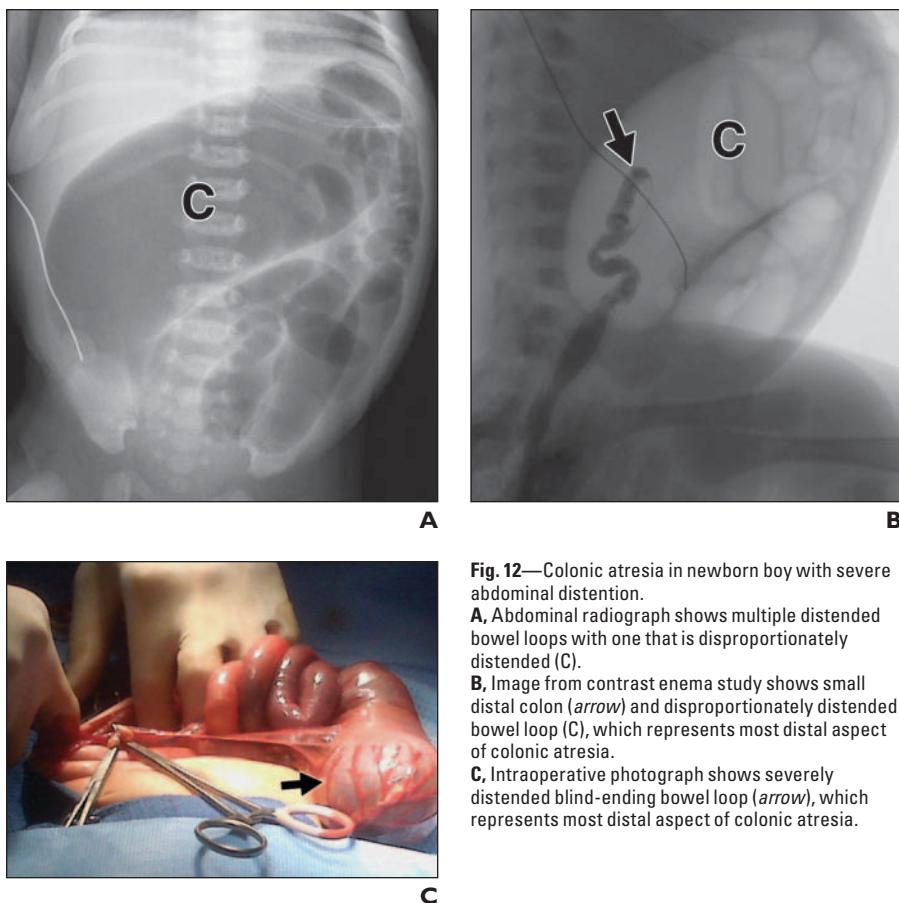


Fig. 12—Colonic atresia in newborn boy with severe abdominal distention.
A, Abdominal radiograph shows multiple distended bowel loops with one that is disproportionately distended (C).
B, Image from contrast enema study shows small distal colon (arrow) and disproportionately distended bowel loop (C), which represents most distal aspect of colonic atresia.
C, Intraoperative photograph shows severely distended blind-ending bowel loop (arrow), which represents most distal aspect of colonic atresia.

Newborns with anal atresia usually present with such signs of lower intestinal obstruction as failure to pass meconium and abdominal distention. Although the diagnosis of anal atresia can be made on the basis of physical examination alone, abdominal radiography can be useful for determining whether the infant has a high or low anal atresia, information that is helpful for surgical planning. Whereas surgical repair for low anal atresia usually involves simple

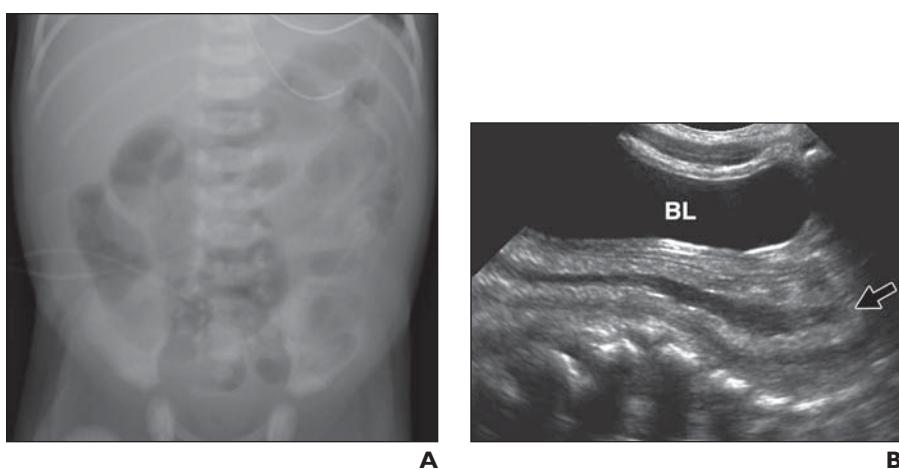


Fig. 13—Anal atresia in newborn boy with abdominal distention and anal atresia detected on physical examination.
A, Frontal abdominal radiograph shows multiple distended bowel loops.
B, Longitudinal ultrasound image through perineum shows fluid-filled blind-ending rectal pouch (arrow). BL = bladder.

anoplasty or dilatation, high anal atresia typically requires a temporizing colostomy with subsequent surgical repair. Abdominal radiographs usually show a low intestinal obstruction pattern (Fig. 13A). A lateral view obtained in the prone position is valuable for showing the level of rectal descent. Ultrasound of the perineum can determine the distance between the perineum and the end of the rectum (Fig. 13B). Further imaging workup for anal atresia includes cystography to assess for associated fistulas with the urinary tract and MRI to completely characterize the anatomy of the pelvic structures.

Conclusion

Intestinal obstructions are the most common surgical emergencies encountered in newborn infants, requiring early and accurate diagnosis. An understanding of the characteristic imaging appearance of various causes of neonatal intestinal obstructions on abdominal radiographs can lead to the correct diagnosis or serve as a guide to the next appropriate step in additional radiologic evaluation. After abdominal radiology that shows the presence of a neonatal high intestinal obstruction, an upper gastrointestinal series is typically performed for further evaluation. However, neonates with classic radiographic findings of high intestinal obstruction, such as duodenal atresia, may directly undergo surgery without any additional imaging study. Similarly, an enema examination is used for further investigation of low intestinal obstruction in neonates.

Suggested Reading

1. Applegate KE, Goske MJ, Pierce G, Murphy D. Situs revisited: imaging of the heterotaxy syndrome. *RadioGraphics* 1999; 19:837–852; discussion, 853–854
2. Berdon WE, Baker DH, Santulli TV, Amoury R. The radiologic evaluation of imperforate anus: an approach correlated with current surgical concepts. *Radiology* 1968; 90:466–471
3. Berdon WE, Slovis TL, Campbell JB, et al. Neonatal small left colon syndrome: its relationship to aganglionosis and meconium plug syndrome. *Radiology* 1977; 125:457–462
4. Berrocal T, Lamas M, Gutierrez J, et al. Congenital anomalies of the small intestine, colon, and rectum. *RadioGraphics* 1999; 19:1219–1236
5. Buonomo C. Neonatal gastrointestinal emergencies. *Radiol Clin North Am* 1997; 35:845–864
6. Donnelly LF. *Fundamentals of pediatric radiology*. Philadelphia, PA: Saunders, 2001
7. Hernanz-Schulman M. Imaging of neonatal gas-trointestinal obstruction. *Radiol Clin North Am* 1999; 37:1163–1186
8. Materne R. The duodenal wind sock sign. *Radiology* 2001; 218:749–750
9. O'Donovan AN, Habra G, Somers S, et al. Diagnosis of Hirschsprung's disease. *AJR* 1996; 167: 517–520
10. Pochaczewsky R, Leonidas JC. The “recto-sigmoid index”: a measurement for the early diagnosis of Hirschsprung's disease. *Am J Roentgenol Radium Ther Nucl Med* 1975; 123:770–777
11. Sato Y, Pringle KC, Bergman RA, et al. Congenital anorectal anomalies: MR imaging. *Radiology* 1988; 168:157–162
12. Stranzinger E, DiPietro MA, Teitelbaum DH, Strouse PJ. Imaging of total colonic Hirschsprung disease. *Pediatr Radiol* 2008; 38:1162–1170
13. Switzer PJ, James CA, Frettag MA. Value and limitations of obstetrical ultrasound uncovering abnormalities at earlier stages. *Can Fam Physician* 1992; 38:121–128