

How I Do It: Imaging Evaluation of Neonatal Bowel Obstructions

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Neonatal bowel obstruction is one of the most common surgical emergencies in neonates in daily clinical practice. Because most causes of bowel obstruction in neonates are different from those in older children and adults, a clear understanding of the specific causes and how to properly evaluate them is important for optimal care. Imaging evaluation plays an essential role in initial workup, accurate diagnosis, and proper guidance of subsequent management in these patients. This up-to-date article provides radiologists with a practical imaging approach and techniques for evaluating neonatal bowel obstruction. The spectrum of underlying causes of neonatal upper and lower bowel obstruction is discussed using a pattern-based approach to analyze characteristic imaging findings. The preferred diagnostic imaging modalities and key imaging features to distinguish these causes of obstructions from other differential diagnoses are highlighted.

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Supplemental material is available for this article.

Neonatal bowel obstruction is the most common surgical emergency in neonates and occurs in approximately one in 2000 births (1,2). Bowel obstructions can be life-threatening, particularly in neonates, and need to be identified accurately and managed appropriately in a timely manner. Most causes of bowel obstruction in neonates differ from those in older children and adults, and understanding the specific causes of neonatal bowel obstructions and how to properly evaluate them is essential (3). In this article, a practical imaging approach and techniques for evaluating congenital neonatal bowel obstruction are discussed. In addition, a pattern-based approach to the spectrum of underlying causes of neonatal bowel obstruction is reviewed. The preferred diagnostic imaging modalities and key imaging features to distinguish these causes of obstructions from other differential diagnoses are highlighted.

Practical Imaging Approach

Neonates with bowel obstruction present with vomiting, abdominal distention, and feeding intolerance, and it can be difficult for surgeons to localize the level of obstruction clinically. A practical pattern-based imaging approach is therefore crucial to localize the obstruction and select the appropriate imaging examination for accurate diagnosis and optimal management (4). Abdominal radiography is typically the initial modality of choice because of its wide availability, portability, and cost-effectiveness. Recognizing normal neonatal bowel gas progression (Fig 1, Appendix S1) helps radiologists distinguish normal findings from obstructive patterns. Radiographs can show signs of bowel obstruction, such as dilated bowel loops, air-fluid levels, static bowel loops, and paucity of bowel gas in the lower abdomen and pelvis. Most neonatal abdominal radiographs are obtained with the patient in a supine position; therefore, air-fluid levels may not be apparent and are not required to diagnose newborn obstruction.

Radiographs are often used to categorize neonatal bowel obstructions into high obstruction, meaning the obstruction is proximal to the ileum, or low obstruction, meaning the obstruction involves the ileum or colon. If fewer than three or four dilated bowel loops are seen, and they are in the upper to mid portion of the abdomen, the bowel obstruction is likely a high obstruction, and a fluoroscopic upper gastrointestinal series (UGI) should be performed (4). If more than four dilated loops are seen, and they are in the abdomen and pelvis, the bowel obstruction is usually low, or distal, and a fluoroscopic contrast enema should be performed for further evaluation (Fig 2).

In some cases, it may be difficult to distinguish the location of the obstruction, and both a UGI and an enema may need to be performed. The decision of which to perform first can be based on the degree of clinical and radiographic suspicion; however, if there is uncertainty, starting with a UGI can be prudent. A UGI can be used to quickly evaluate for a potentially life-threatening upper tract obstruction, such as malrotation with midgut volvulus, while avoiding obscuration of the duodenum by contrast material in the colon. If the UGI is normal, an enema can then be performed to evaluate for lower tract obstruction.

If bowel perforation is suspected, cross-table lateral, decubitus, or upright radiography increases the sensitivity for detecting pneumoperitoneum (5–7). Radiographic signs of pneumoperitoneum include lucency over the liver with outlining of the falciform ligament (football sign), visualization of both sides of bowel walls (Rigler sign), and triangular lucencies between bowel loops (telltale triangle sign) (Fig 3).

Some causes of bowel obstruction can be diagnosed from the radiograph alone, such as esophageal or duodenal atresia, occasionally jejunal atresia, and inguinal hernia. Finally, if an obstruction is suspected, evaluation and care of the neonate should occur in a center with pediatric specialists, including pediatric surgeons, pediatric radiologists, and neonatologists.

Abbreviation

UGI = upper gastrointestinal series

Summary

A practical patterned approach to neonatal bowel obstructions helps radiologists localize the obstruction and perform the appropriate imaging examination to accurately make the diagnosis and help guide proper management.

Essentials

- Most causes of bowel obstruction in neonates differ from those in older children and adults, and knowing the common causes of neonatal bowel obstruction is essential for accurate diagnosis and optimal management.
- In suspected neonatal bowel obstruction, abdominal radiography is typically the preferred initial imaging modality because of its wide availability, portability, and cost-effectiveness.
- Abdominal radiography can be used to look for signs of bowel obstruction or perforation, distinguish between upper and lower intestinal tract obstructions, and guide next steps in management, such as further imaging.
- Most upper bowel obstructions can be evaluated with a fluoroscopic upper gastrointestinal series, and findings generally fit one of six patterns: normal, duodenal web, duodenal stenosis or annular pancreas, malrotation, malrotation with midgut volvulus, and duodenal obstruction.
- Most lower bowel obstructions can be evaluated with a fluoroscopic contrast enema, and findings generally fit one of four patterns: abrupt caliber change (associated with classic Hirschsprung disease), small caliber of the left colon to near the level of the splenic flexure (seen with small left colon syndrome), diffuse microcolon, and short microcolon.

High Bowel Obstruction

Most high obstructions in neonates are related to one of six anomalies: esophageal atresia, duodenal atresia, duodenal stenosis with or without annular pancreas, duodenal web, malrotation with midgut volvulus, or jejunal atresia. Each of these is discussed in this section, along with the preferred diagnostic imaging modalities and key imaging features. Although esophageal, duodenal, and occasionally jejunal atresia can be detected and diagnosed using radiography alone, the others typically require UGI for accurate diagnosis. There are six general patterns of UGI that radiologists should be familiar with, to aid in making the correct diagnosis (3) (Table 1, Fig 4). Normal duodenal variants have been previously described and can make interpreting UGI studies more challenging for radiologists not familiar with them (8); however, these variants generally still fit within the six patterns. The technique and contrast agent considerations for performing a UGI have been previously described (9–12) and are summarized in Appendix S1.

UGI Pattern 1: Normal

Pattern 1 represents a normal examination with a smooth contour of the stomach and duodenum without filling defects, narrowing, or dilatation, with free flow of contrast material from the stomach to the duodenojejunal junction at the ligament of Treitz. The duodenum must also have the normal retroperitoneal course (best seen on the lateral view), should cross the midline (best seen on the frontal view), and should ascend to the left of the spine to near the same level as the duodenal bulb (Fig 4A). These UGI findings demonstrate normal rotation of the bowel and help exclude malrotation or duodenal obstruction.

UGI Pattern 2: Duodenal Web

Pattern 2 involves normal rotation and position of the duodenojejunal junction but with dilatation of the proximal duodenum and, typically, focal rounding and dilatation of the transverse duodenum. An often-seen curvilinear lucency at the distal edge of the dilatation represents an intraluminal fenestrated web or membrane, which partially obstructs the lumen. This partial occlusion is thought to be caused by incomplete luminal recanalization (13). On radiographs, it can be distinguished from duodenal atresia by the presence of distal bowel gas. A fluoroscopic UGI can help distinguish a luminal web from other causes of upper tract obstruction and can show the classic windsock sign, caused by contrast outlining the thin web or membrane (Fig 4B). With any partial duodenal obstruction, the flow of the contrast material should be followed to the jejunum to document normal left upper quadrant position of the duodenojejunal junction and exclude malrotation with midgut volvulus.

UGI Pattern 3: Duodenal Stenosis or Annular Pancreas

Pattern 3 shows normal rotation and position of the duodenojejunal junction but with dilatation of the proximal duodenum and focal narrowing, usually centered in the descending duodenum. Duodenal stenosis, annular pancreas, and duodenal web can look similar radiographically and on UGI images, so distinguishing them before surgical intervention may not be possible.

Duodenal stenosis may manifest with or without an annular pancreas. Stenosis is caused by partial atresia of a duodenal segment (leading to stenosis) and/or by an abnormal ring of pancreatic tissue circumscribing the descending duodenum (causing luminal narrowing). Radiographically, duodenal stenosis often manifests as a dilated stomach and proximal duodenum, but because obstruction is incomplete, distal bowel gas will be present. Fluoroscopic UGI typically shows the focal narrowing of the duodenum (Fig 4C). CT or MRI may be necessary to evaluate the narrowing and assess any extrinsic causes, such as a ring of pancreatic tissue around the duodenum.

UGI Pattern 4: Malrotation

Pattern 4 shows abnormal positioning of the duodenojejunal junction (Fig 4A). The duodenum does not cross the midline and typically has a more anterior course in the lateral position. The jejunum tends to be mainly located on the right side of the abdomen (Fig 4D). Although malrotation itself does not constitute a bowel obstruction or surgical emergency, it predisposes patients to developing midgut volvulus. The sensitivity and specificity of UGI for malrotation have been reported to be 93%–100% and 64%–100%, respectively (14).

Malrotation occurs during embryonic development when the bowel fails to complete its normal 270° counterclockwise rotation. The resulting abnormal positioning allows the bowel to move more freely and potentially twist around its own mesentery and vascular supply. Abnormal fibrous peritoneal bands, or Ladd bands, may also be present in malrotation and may cause duodenal narrowing or obstruction. Patients with malrotation can be asymptomatic or have intermittent abdominal pain if the malrotation is associated with intermittent volvulus causing bowel obstruction.

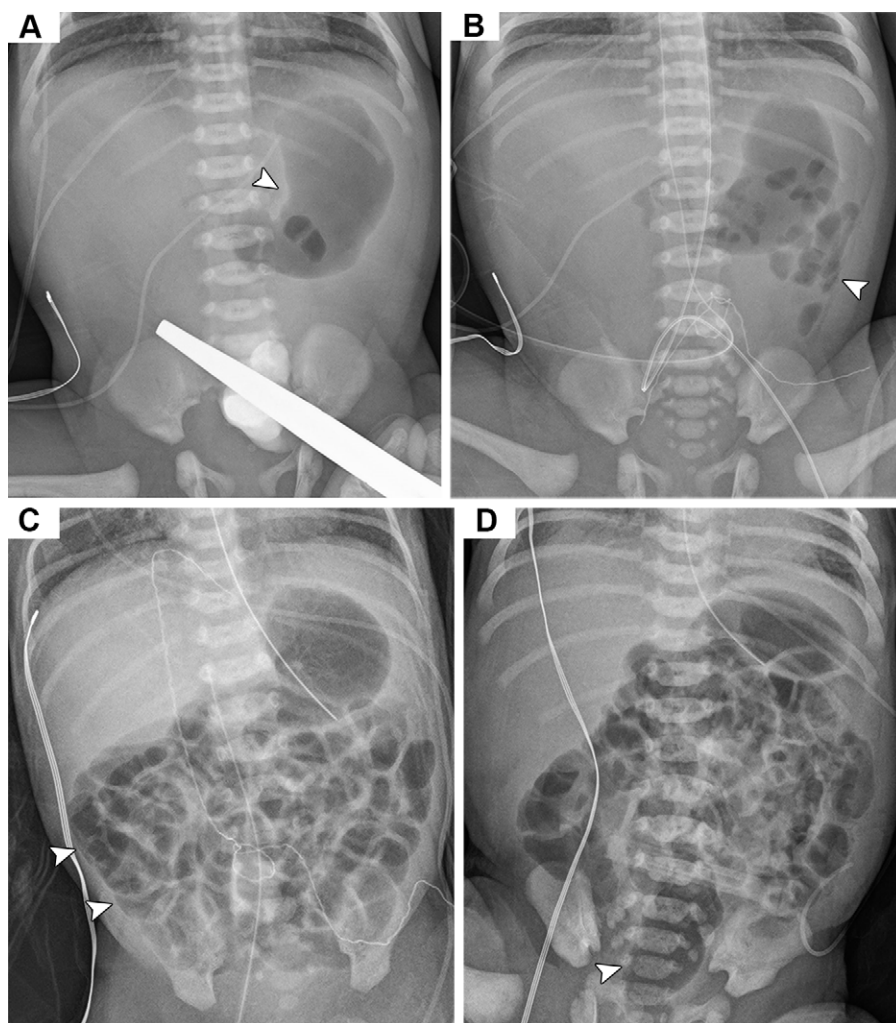


Figure 1: Frontal abdominal radiographs show normal progression of bowel gas in a neonate during the 1st day of life. **(A)** At approximately 1 hour of life, gas should fill the stomach (arrowhead). **(B)** By 3 hours, small bowel loops should fill with air (arrowhead). **(C)** Around 5 hours, bowel gas should arrive in the proximal colon (arrowheads). **(D)** Finally, between 12 and 24 hours of life, gas should reach the rectum (arrowhead).

UGI Patterns 5 and 6: Malrotation with Midgut Volvulus and Duodenal Obstruction

Pattern 5 shows signs of malrotation but with the addition of the classic “corkscrew” appearance of narrowed bowel loops coiled as the bowel twists on its mesentery (Fig 4E). Pattern 6 shows the flow of contrast material abruptly stopping in the duodenum, which is highly concerning for midgut volvulus with obstruction (Fig 4F). Patients with midgut volvulus often develop bilious emesis. Although midgut volvulus may occur at any age, up to 90% of symptomatic patients present in the 1st year of life (14).

Radiography is insensitive to malrotation with midgut volvulus and should not be relied on to exclude this entity. Radiographs may even be normal in this setting. If midgut volvulus is suspected, or if a neonate presents with bilious emesis, emergent imaging with US or UGI (depending on the institution) should be performed immediately. Delayed diagnosis can lead to infarction of the small bowel and potentially short bowel syndrome or death.

UGI for the diagnosis of malrotation with midgut volvulus is reported to have a sensitivity of 54%–79% and specificity of

98% (14). Although UGI is conventionally the imaging test of choice, some institutions use Doppler US to search for signs of malrotation and midgut volvulus. These signs include a reversed anatomic relationship of the superior mesenteric artery and vein (which indicates malrotation) and a swirling of the mesenteric vessels in a whirlpool sign (which indicates midgut volvulus) (Fig 5). US is reported to have a sensitivity and specificity for midgut volvulus of 83%–100% and 91%–100%, respectively (14). The advantages and disadvantages of UGI versus US for this diagnosis have been compared previously (14). Each practice and institution might approach the diagnostic imaging of this condition differently based on staff training and expertise. Often, if US is used and is inconclusive for midgut volvulus, emergent UGI is then performed. Treatment of midgut volvulus is emergent surgery.

Other Causes of High Bowel Obstruction

Three other causes of neonatal high bowel obstruction are esophageal atresia, duodenal atresia, and jejunal atresia or stenosis (Fig 6). However, these causes often do not require UGI for diagnosis. These may be differentiated from other diagnoses

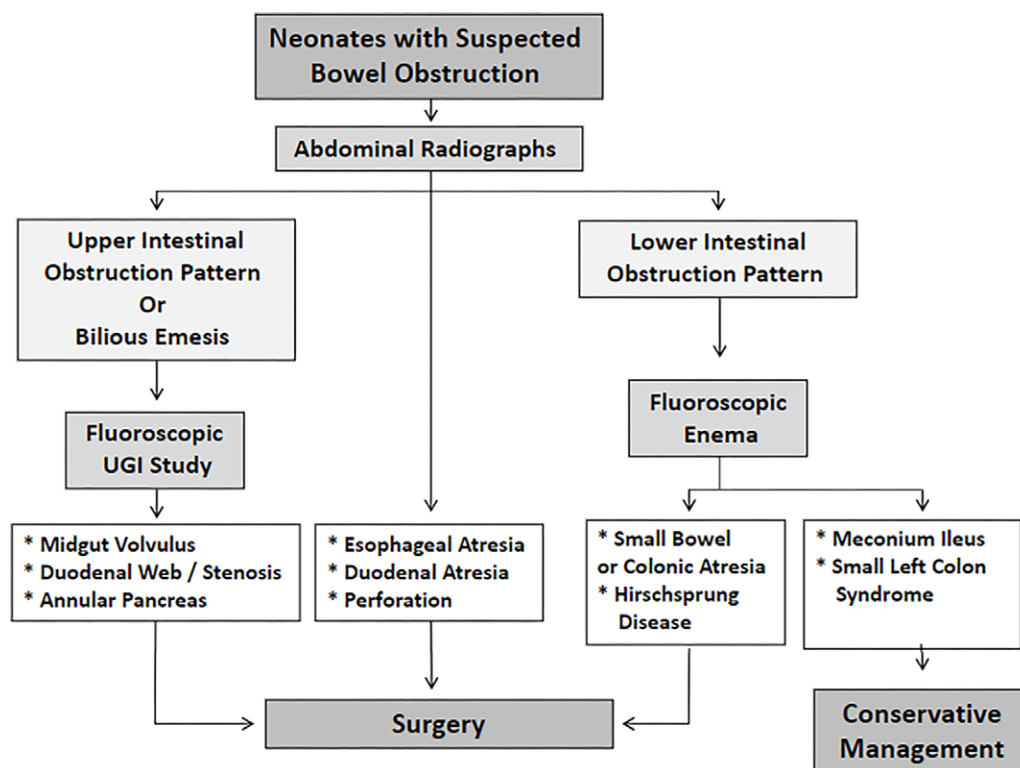


Figure 2: Practical imaging approach for neonates with suspected bowel obstruction. Typically, the presence of fewer than three or four dilated bowel loops located in the upper to mid portion of the abdomen indicates a high bowel obstruction, and a fluoroscopic upper gastrointestinal series (UGI) should be performed. The presence of more than four dilated loops in the abdomen and pelvis typically indicates a low, or distal, bowel obstruction, in which case a fluoroscopic contrast enema should be performed. (Adapted, with permission, from reference 3.)

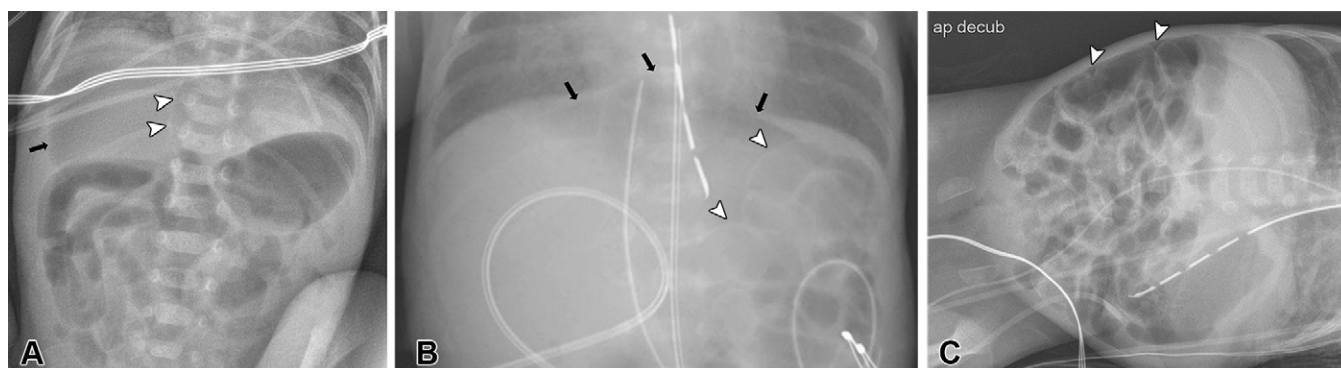


Figure 3: Radiographic signs of free air. **(A)** Supine frontal radiograph in a premature infant with free intraperitoneal air over the liver (arrow) and outlining of the falciform ligament (arrowheads), known as the football sign. **(B)** Frontal supine radiograph in a different premature infant with free intraperitoneal air around the central diaphragm (arrows), with visualization of both sides of the wall of bowel loops in the left upper abdomen (arrowheads), known as the Rigler sign. **(C)** Right-side-up lateral decubitus radiograph in a different neonate shows small triangular locules of free peritoneal air (arrowheads), known as the telltale triangle sign.

with radiographic findings alone, or with the combination of radiographic findings and supporting clinical information.

Esophageal Atresia

Esophageal atresia occurs when there is abnormal embryologic development of the longitudinal ridge separating the trachea and the esophagus. This condition occurs within a spectrum often classified based on the presence of a fistulous connection between the esophagus and trachea. Esophageal atresia can be associated with VACTERL association (vertebral anomalies, anorectal atresia, cardiac abnormality, tracheoesophageal fistula, renal anomaly, and limb defect) (15).

Affected patients may present with sialorrhea, choking, or respiratory distress shortly after birth. Prenatal imaging with US or MRI may depict polyhydramnios, esophageal pouch, or small stomach. Postnatal radiography may show failure of an enteric tube to pass below the upper to mid esophagus (Fig 6A). If esophageal atresia is present without a fistula to the airway, bowel gas is absent, whereas the presence of bowel gas suggests that a fistula with the airway is present. Timing of surgical repair may vary based on the length of the esophageal atretic segment, but repair often occurs in the 1st week of life. Fluoroscopic contrast esophagography is not indicated before surgery because of the risk of aspiration. However, after repair,

Table 1. Neonatal High Bowel Obstructions

Pattern and Diagnosis	Diagnostic Examination	Distinguishing Features
UGI pattern 1: normal	UGI	Retroperitoneal course of the duodenum with duodenojejunal junction to the left of the spine at the level of the duodenal bulb; no dilatation or narrowing
UGI pattern 2: duodenal web	UGI	Dilated proximal duodenum with outline of thin web or membrane, known as the windsock sign
UGI pattern 3: duodenal stenosis or annular pancreas	UGI as first-line imaging study; CT or MRI may be needed for evaluating annular pancreas	Focal narrowing of the descending duodenum
UGI pattern 4: malrotation	UGI as first-line imaging study; US, CT, or MRI can also show abnormal findings	Abnormal positioning and course of the duodenum
UGI pattern 5: malrotation with midgut volvulus	UGI or US as first-line imaging study (institution dependent)	Corkscrew configuration of the duodenum and jejunum on UGI images; reversed anatomic relationship of the superior mesenteric artery and vein, with swirling of the mesenteric vessels at US
UGI pattern 6: duodenal obstruction	UGI; US can be used if midgut volvulus suspected	Abrupt cutoff of contrast near the transverse duodenum on UGI images
Other causes of high bowel obstruction		
Esophageal atresia	Babygram (chest and abdominal radiography) after attempted enteric tube placement	Failure to pass nasogastric tube into distal esophagus or stomach; dilated air-filled upper thoracic pouch or esophagus; other VACTERL anomalies
Duodenal atresia	Abdominal radiography	Dilated, air-filled stomach and proximal duodenum (double bubble sign) with absent distal bowel gas
Jejunal atresia or stenosis	Abdominal radiography and contrast enema	Dilated proximal small bowel loops, with paucity of distal bowel gas in the pelvis; microcolon at contrast enema if jejunal atresia is present

Note.—UGI = upper gastrointestinal series, VACTERL = vertebral anomalies, anorectal atresia, cardiac abnormality, tracheoesophageal fistula, renal anomaly, and limb defect.

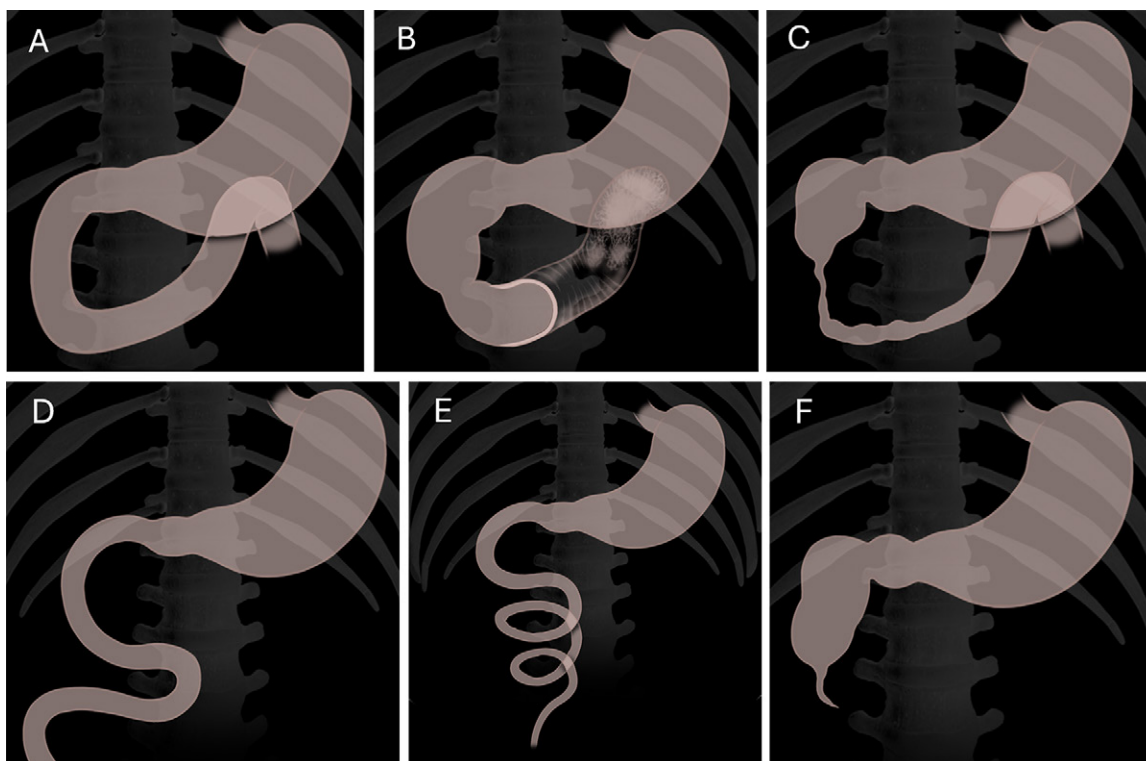


Figure 4: Upper gastrointestinal series (UGI) patterns for neonatal high bowel obstructions. **(A)** Normal UGI pattern. **(B)** Duodenal web, showing the classic windsock sign caused by contrast material outlining the thin web or membrane. **(C)** Duodenal stenosis or annular pancreas, showing focal narrowing of the duodenum. **(D)** Malrotation. **(E)** Malrotation with midgut volvulus with corkscrew configuration. **(F)** Duodenal obstruction, which can be seen with midgut volvulus. (Adapted, with permission, from reference 4.)

esophagography may be used to assess for anastomotic stenosis, leak, or recurrent fistula (16).

Duodenal Atresia

Duodenal atresia results from failure of recanalization of the lumen of the duodenum. Up to 30% of patients with duodenal atresia have trisomy 21 (Down syndrome) (13,17). The diagnosis may be made prenatally with US or MRI, which shows a dilated stomach and proximal duodenum. Postnatal radiographs classically show the double bubble sign (dilated, air-filled stomach and proximal duodenum) (Fig 6B) with absent distal bowel gas, and this sign is usually sufficient for diagnosis. UGI is not needed for the diagnosis.

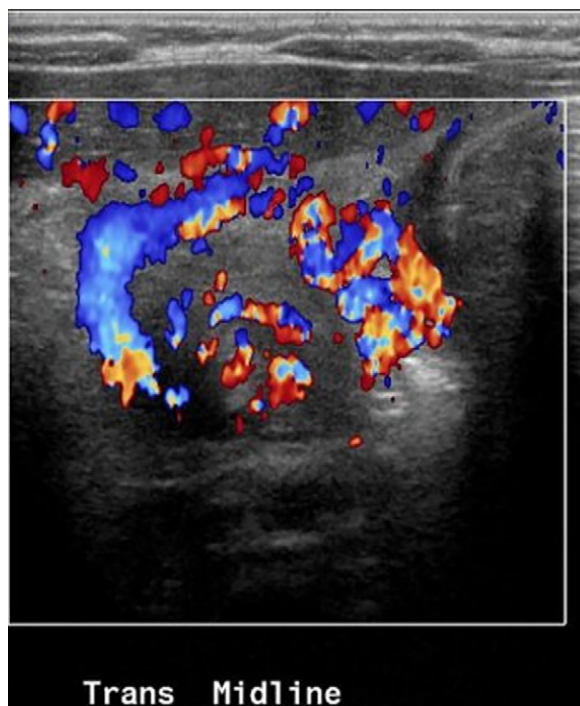


Figure 5: Sonographic sign of midgut volvulus. Transverse color Doppler US image of the abdomen near the midline shows twisting of the superior mesenteric artery and vein in the whirlpool sign of midgut volvulus. (Reprinted, with permission, from reference 3.)

Jejunal Atresia or Stenosis

Neonatal obstruction of the jejunum is generally related to in utero vascular insults causing congenital atresia or stenosis. Multiple areas of atresia often coexist. Abdominal radiographs may show a few dilated bowel loops, or even the triple bubble sign (Fig 6C) if a proximal jejunal atresia is present (2,11), but these features are not always seen because of decompression via vomiting or suction. Fluoroscopic UGI is frequently needed to confirm the diagnosis and to exclude malrotation with midgut volvulus. In addition, a contrast enema can be performed to exclude meconium ileus. Contrast enema may show a diffuse microcolon and can suggest additional sites of distal small bowel atresia. If the jejunal atresia or stenosis is proximal, contrast enema findings may still be normal because the distal jejunum and ileum can produce enough secretions to create sufficient meconium to enter the colon; however, persistent dilated proximal small bowel loops will remain, with obstructive symptoms.

Low Bowel Obstruction

Low, or distal, bowel obstructions in neonates may be caused by six main congenital abnormalities: ileal atresia, meconium ileus, small left colon syndrome, Hirschsprung disease, colonic or anal atresia, and megacystis-microcolon-intestinal hypoperistalsis syndrome. Most causes should be evaluated with a contrast enema and will generally fit within four imaging patterns (4) (Table 2, Fig 7). Knowing these four patterns can help narrow the differential diagnosis and guide management. The technique for performing contrast enemas is described in Appendix S1 (18).

Enema Pattern 1: Classic Hirschsprung Disease

Pattern 1 shows an abrupt caliber change of the colon, usually involving the rectosigmoid, with a rectosigmoid ratio less than one, and is typically due to classic Hirschsprung disease. Irregular colonic contractions can sometimes be seen (3).

Hirschsprung disease accounts for approximately 15%–20% of bowel obstructions in neonates (17), and about 2% of patients with Hirschsprung disease also have trisomy 21 (Down syndrome). Hirschsprung disease is caused by failure of normal migration of colonic ganglion cells from the proximal to the distal

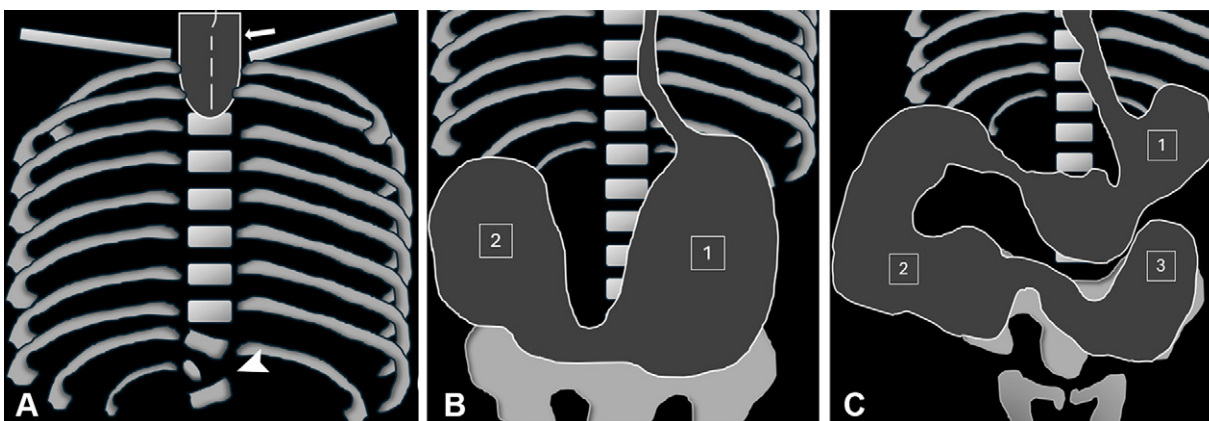


Figure 6: Other causes of high bowel obstructions in neonates. **(A)** Esophageal atresia, with an enteric tube stuck in an air-distended esophageal pouch (arrow) and vertebral anomalies (arrowhead). **(B)** Duodenal atresia, with the double bubble sign (dilated stomach [area 1] and duodenum [area 2]) with no distal bowel gas. **(C)** Jejunal atresia, with the triple bubble sign (air-distended stomach [area 1], duodenum [area 2], and proximal jejunum [area 3]) with no distal bowel gas.

Table 2. Neonatal Low Bowel Obstructions

Pattern and Diagnosis	Diagnostic Examination	Distinguishing Features
Enema pattern 1: classic Hirschsprung disease (most likely diagnosis)	Contrast enema	Abrupt caliber change usually involving the rectosigmoid junction, with rectosigmoid ratio < 1
Enema pattern 2: small left colon syndrome	Contrast enema	Smaller-caliber left colon to near the level of the splenic flexure, with meconium filling defects in the left colon
Enema pattern 3: diffuse microcolon	Contrast enema	
Ileal atresia	Contrast enema	Diffuse microcolon with failure to reflux contrast material past atretic distal ileum; attempting to reflux contrast material is not always practically possible, and other causes of diffuse microcolon should remain in the differential diagnosis
Meconium ileus	Contrast enema	Diffuse microcolon with small filling defects seen in the ileocecal region from impacted meconium
Total colonic Hirschsprung disease	Contrast enema	Diffuse microcolon; rectal suction biopsy may be needed to distinguish this from other entities
Megacystis-microcolon-intestinal hypoperistalsis syndrome	Contrast enema for bowel obstruction workup; US and/or voiding cystourethrography for urinary workup	Diffuse microcolon plus dilated urinary tract and bladder
Enema pattern 4: short microcolon	Contrast enema	Short microcolon
Other causes of low bowel obstruction		
Anorectal malformations	US, voiding cystourethrography, and/or MRI	Blind-ending anus or rectum; fistulous connection of the colon to other pelvic structures in high anorectal malformations
Congenital inguinal hernia	Radiography and/or US	Bowel loops within the inguinal canal and/or scrotum

colon. This results in the affected colonic segment being unable to relax, leading to a functional obstruction. The affected aganglionic segment starts at the anus and is continuous with any portion of the colon affected proximally because of the craniocaudal cell migration of the colonic ganglia (19). Affected segments may include the rectosigmoid colon (most common, approximately 75% of patients), longer segments, or the entire colon (approximately 17% of patients) (20–22) (Fig 7A). Affected patients may present with delayed passage of meconium, infrequent stooling, and abdominal distention.

Contrast enema for Hirschsprung diagnosis has a mean sensitivity and specificity of 70% and 83%, respectively (22). Therefore, if Hirschsprung disease is clinically suspected, even in the case of a normal or nondiagnostic contrast enema, rectal suction biopsy should be performed for definitive diagnosis. In patients with Hirschsprung disease, the rectosigmoid ratio is usually observed to be less than one during the contrast enema; however, if the transition point is proximal to the sigmoid colon, the rectosigmoid ratio may not measure less than one because the entire rectosigmoid segment lacks ganglion cells. Hirschsprung disease may also manifest with a diffuse microcolon (enema pattern 3), but this is less common than the classic presentation. An ultrashort segment form of Hirschsprung disease has also been described but is rare and is identified at manometry.

Enema Pattern 2: Small Left Colon Syndrome

Pattern 2 shows a normal rectosigmoid ratio, but with a relatively small caliber of the left colon to near the level of the splenic flexure. The small left colon often contains small filling defects representing meconium.

Small left colon syndrome, also known as functional immaturity of the colon or meconium plug syndrome, is usually a

self-limited functional obstruction in neonates; it is thought to be caused by immaturity of the ganglion cells in the myenteric nerve plexus of the colon. Small left colon syndrome is the most common cause of infants' not passing meconium within the first 2 days of life (13). Small left colon syndrome is frequently associated with patients' mothers having diabetes or being given magnesium sulfate for preeclampsia during pregnancy. Contrast enema often shows a relatively small caliber of the left colon to the region of the splenic flexure, with multiple intraluminal filling defects from meconium (Fig 7B). The rectum, ascending, and transverse colon are usually normal caliber, although the more proximal colon can be slightly dilated because of the functional obstruction of the left colon. Contrast enema can be both diagnostic and therapeutic because the contrast material can help the infant expel the meconium and resolve the obstruction. If symptoms do not resolve quickly, Hirschsprung disease should be excluded with a rectal wall biopsy (23).

Enema Pattern 3: Diffuse Microcolon

Pattern 3 is distinguishable in that the entire colon is very small in caliber (because of failure to pass meconium into the colon) (Fig 7C). Unlike the two previous enema patterns, this pattern is not specific to one disease entity and should prompt a differential diagnosis to determine the correct diagnosis. The four main diagnoses associated with this pattern are discussed here. Some have distinguishing features (not always visible), which can help narrow the differential diagnosis and guide subsequent management.

Ileal Atresia

Ileal atresia is thought to result from intrauterine ischemia, similar to jejunal atresia. Patients may present with failure to

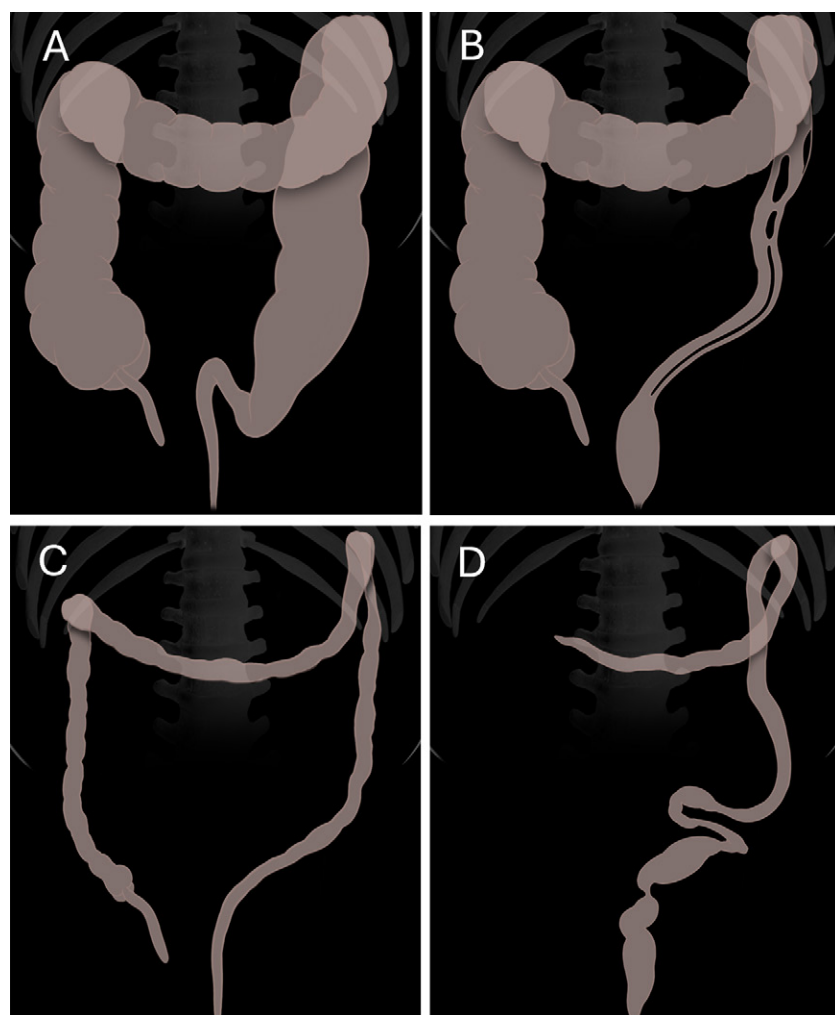


Figure 7: Contrast enema patterns for neonatal low bowel obstructions. **(A)** Classic Hirschsprung disease, with abrupt caliber change involving the rectosigmoid colon. **(B)** Small left colon syndrome, with smaller-caliber colon to the splenic flexure and multiple meconium filling defects. **(C)** Diffuse microcolon. This pattern can be seen in meconium ileus, small bowel atresia, or, less commonly, total colonic Hirschsprung disease or megacystis-microcolon–intestinal hypoperistalsis syndrome. **(D)** Short microcolon, seen with colonic atresia. (Adapted, with permission, from reference 4.)

pass meconium, abdominal distention, and vomiting. Contrast enema usually shows a diffuse microcolon, in which case an attempt to reflux contrast material into the ileum is often helpful as contrast material may not be able to be refluxed past an atretic segment of distal ileum. This may help distinguish this entity from others. Ileal atresia should always be considered when there is a microcolon and disproportionately marked dilatation of small bowel loops.

Meconium Ileus

Meconium ileus is caused by impaction of desiccated meconium in the terminal ileum near the ileocecal valve, with resultant obstruction and failure of meconium migration into the colon. It is usually associated with cystic fibrosis and may be the first clinical manifestation of the disease (13). Contrast enema usually shows a diffuse, unused microcolon. If contrast material can be refluxed into the terminal ileum, small filling defects (impacted meconium pellets) may be seen, which can help differentiate this entity from other causes of diffuse microcolon. Patients with this finding should undergo testing for cystic fibrosis.

Total Colonic Hirschsprung Disease

As discussed earlier, Hirschsprung disease is caused by failure of migration of ganglion cells within the colon. If this failure of migration affects the entire colon, rather than just a distal segment, it can cause a diffuse microcolon pattern at contrast enema.

Megacystis-Microcolon–Intestinal Hypoperistalsis Syndrome

Megacystis-microcolon–intestinal hypoperistalsis syndrome is a very rare cause of functional bowel obstruction in neonates. It may be associated with malrotation and can lead to death (24–26). The cause of this condition is not fully understood. However, abnormalities of the ganglion cells associated with smooth muscle may explain why this condition manifests with absent bowel peristalsis and dilated urinary collecting systems and bladder (25). Contrast enema typically shows a diffuse microcolon due to meconium not traversing through the small bowel because of the lack of peristaltic movement. This disease should be included in the differential diagnosis when there is failure to pass meconium and a diffuse microcolon is seen at contrast enema. Dilatation of the urinary system should help make this diagnosis.

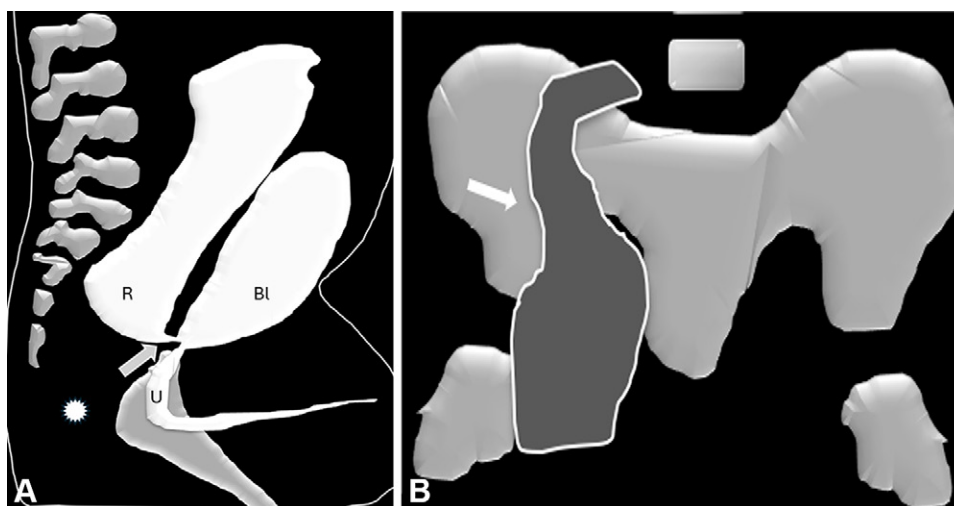


Figure 8: Other causes of low bowel obstructions in neonates. **(A)** Anorectal malformation, with a blind-ending rectum (R) with a fistulous connection (arrow) to the bladder (Bl) neck, just superior to the urethra (U). There is no rectum in the expected presacral space (*). **(B)** Congenital inguinal hernia, with air-distended bowel loop in the right inguinal canal (arrow).

Enema Pattern 4: Short Microcolon

Pattern 4 is similar to pattern 3 in that the colon is diffusely small in caliber, but in pattern 4 the colon is also short in length, with failure of contrast material to reflux into the more proximal colon (Fig 7D). Short microcolon is seen with colonic atresia, which accounts for about 2%–15% of all intestinal atresias and is less common than small bowel atresias (13,27). Colonic atresia most commonly occurs proximally to the splenic flexure.

Other Causes of Low Intestinal Obstruction

Two other causes of neonatal low intestinal obstruction are anorectal malformations and congenital inguinal hernia. However, these conditions often do not require contrast enema for diagnosis. They can sometimes be diagnosed with radiography alone or with the combination of radiography and supporting clinical information.

Anorectal Malformations

Anorectal malformation is an umbrella term that encompasses a spectrum of distal bowel abnormalities, including atresia (anal or rectal) and anal stenosis. Patients with these abnormalities may present with signs of a lower tract obstruction and failure to pass meconium. Associated pathologic abnormalities can co-occur, as seen with the Currarino triad (anorectal malformation, malformed sacrum, and presacral mass) and VACTERL association. Anorectal malformations are generally classified as high lesions or low lesions based on the position of the blind-ending rectum relative to the levator ani muscle (28). In high lesions, the blind-ending rectum is located in the pelvis some distance from the skin and often has a fistulous connection to another pelvic organ, such as the urethra, bladder, or vagina (Fig 8A). In low lesions, the blind-ending rectum is closer to the skin, and these lesions tend not to have fistulous connections.

US can help show the blind pouch and its distance to the skin, as well as other abnormalities. MRI can show additional intrapelvic abnormalities and help the radiologist visualize the sphincter complex in relation to other organs. Fluoroscopic voiding cystourethrography may help identify fistulas.

Congenital Inguinal Hernia

Congenital inguinal hernias are estimated to occur in approximately 1%–2% of children, and up to 10% of hernias are complicated by acute bowel obstruction (29). Many of these cases are found clinically during physical examination. Abdominal radiography can show air-filled bowel loops within the inguinal canal or scrotum (Fig 8B), as well as signs of obstruction, if present. In some cases, focused US can help the radiologist visualize herniated bowel loops and abdominal contents and estimate the size of the fascial defect.

Conclusion

Radiologists frequently encounter neonates with bowel obstructions, which can be life-threatening. In neonates with clinically suspected bowel obstruction, abdominal radiography is the first-line imaging modality to evaluate for diagnoses that can be made radiographically, to assess for signs of bowel obstruction or perforation, and to help guide further imaging of the upper or lower gastrointestinal tract. An understanding of practical imaging techniques and of the characteristic imaging findings of underlying causes of neonatal bowel obstructions based on a pattern-based approach has great potential for aiding accurate and timely diagnosis. This, in turn, can lead to optimal management of obstructions.

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