

# Ultrasound for Midgut Malrotation and Midgut Volvulus: *AJR* Expert Panel Narrative Review

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Pediatric Imaging • *AJR* Expert Panel Narrative Review

## Keywords

bowel, children, emergency, midgut malrotation, midgut volvulus, neonatal, obstruction, pediatric, surgery, upper gastrointestinal (UGI)

Submitted: Dec 15, 2021

Revision requested: Dec 23, 2021

Revision received: Jan 20, 2022

Accepted: Jan 24, 2022

First published online: Feb 2, 2022

The authors declare that they have no disclosures relevant to the subject matter of this article.

An electronic supplement is available online at [doi.org/10.2214/AJR.21.27242](https://doi.org/10.2214/AJR.21.27242).

Midgut volvulus in association with malrotation is a pediatric surgical emergency. Prompt and accurate diagnosis is necessary to avoid bowel ischemia and necrosis, thereby reducing morbidity and mortality. Historically, the fluoroscopic upper gastrointestinal series has been the preferred imaging modality for the evaluation of both midgut malrotation and volvulus, although the use of ultrasound (US) is increasing. In this Narrative Review, we describe the findings of midgut malrotation and volvulus on US, including practical tips for acquisition and interpretation; discuss the advantages and challenges of both imaging modalities; and propose a path and safeguards for possible transition to the use of US as the first-line modality for diagnosis based on our experience in imaging children with midgut malrotation and volvulus.

Midgut malrotation (hereafter, referred to as “malrotation”) is a developmental rotation and fixation abnormality of the embryonic bowel, resulting in a narrow mesenteric root attachment. Malrotation may be associated with bowel obstruction from Ladd bands or midgut volvulus (hereafter, referred to as “volvulus”) at any age, occurring most commonly in early infancy, although occasionally occurring in older patients with chronic symptoms [1–4]. In volvulus, the bowel twists around the superior mesenteric artery (SMA) and narrow mesenteric root, leading to bowel obstruction and possible infarction that results in substantial morbidity and mortality if not emergently diagnosed and surgically corrected [5]. For several decades, the fluoroscopic upper gastrointestinal (UGI) series has been the preferred imaging modality for the diagnosis of malrotation and volvulus [6–8]. More recently, a growing body of literature supports the use of ultrasound (US) to supplement or replace the UGI series for the investigation of both conditions [9–32]. In this Narrative Review, we describe the findings of malrotation and volvulus on US, including practical tips for acquisition and interpretation; discuss the advantages and challenges of US and the UGI series; and propose a path and safeguards for possible transition to the use of US as the first-line modality for diagnosis based on our experience in imaging children with midgut malrotation and volvulus.

## Diagnosing Malrotation and Volvulus With Ultrasound

Normal rotation is established on US when the superior mesenteric vein (SMV) courses to the right of an axis formed between the SMA and the aorta (i.e., the SMA-aorta axis) and a retroperitoneal (i.e., retromesenteric) third portion of the duodenum (hereafter, referred to as “D3”) is unequivocally identified between the SMA and aorta (Fig. 1). It is paramount to show continuity of the duodenum from the pylorus to D3 to avoid confusing D3 with jejunal loops that may also course between the SMA and aorta.

Because radiologists may be unfamiliar with identifying D3 on US, it is recommended to start by identifying the pylorus and duodenal bulb, as is done for an US examination performed to assess for pyloric stenosis. Subsequently, the probe is moved or fanned caudally to visualize

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[doi.org/10.2214/AJR.21.27242](https://doi.org/10.2214/AJR.21.27242)

*AJR* 2022; 218:931–939

ISSN-L 0361–803X/22/2186–931

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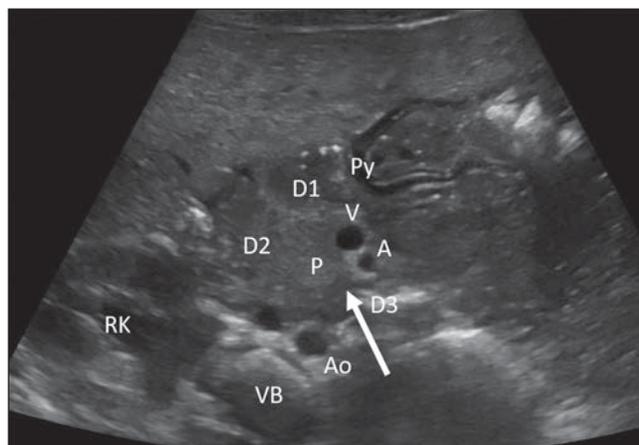
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the second portion of the duodenum (hereafter, referred to as “D2”), which courses rightward of the pancreatic head (Fig. 1). D2 can then be connected to D3, which is the horizontal continuation of the duodenum just below the pancreas. In general, if the kidneys (when orthotopic) are not present on the same view in which the suspected D3 position is visualized, then the finding may represent a jejunal loop rather than D3. Some groups advocate for administration of normal saline or sterile water by an enteric tube to better delineate the duodenal course, using the fluid-filled stomach as a sonographic window and following the bolus of fluid through the duodenum [10, 15, 20, 26, 32]. Intraluminal air can obscure the bowel on US, a limitation that is exacerbated by aerophagia related to crying. Providing a comforting environment for the child, such as by scanning with the patient in the caregiver’s arms, may also increase the success rate by decreasing aerophagia and motion from crying. Turning the patient into a right lateral decubitus position may displace the air and allow emptying of gastric contents into the duodenum. Graded compression, similar to that which is advised for appendiceal US, is also helpful. The use of a small-footprint microconvex probe (Video S1, which can be viewed in the electronic supplement to this article at doi.org/10.2214/AJR.21.27242) allows graded compression without interference from the anterior ribs, in contrast to the use of larger-footprint linear probes.

Findings on US that are reported to suggest the presence of malrotation include an SMV located anterior to or to the left of the SMA-aorta axis, an intraperitoneal course of D3 anterior to the SMA (rather than its normal retroperitoneal course between the aorta and the SMA), abnormal cecal position (or only small bowel in the right lower quadrant), and a hypoplastic pancreatic

uncinate process (not extending posterior to the superior mesenteric vessels) [10, 15, 17, 20, 22, 24, 28, 32–36] (Fig. 2 and Video S2, the latter of which can be viewed in the electronic supplement to this article at doi.org/10.2214/AJR.21.27242). The sensitivity and specificity of these findings on US vary across studies (Table 1).



**Fig. 1**—Normal midgut rotation in 1-month-old girl with emesis. Transverse gray-scale ultrasound image at level of pancreas (P) shows normal SMV-SMA-aorta axis relationship with SMV (V) coursing anterior and to right of SMA (A), which has echogenic collar. First (D1), second (D2), and third (D3) portions of duodenum are normal. D3 is echogenic due to intraluminal air and courses in retroperitoneal fashion between SMA and aorta (Ao) at level of pancreas and right kidney (RK). Uncinate process of pancreas (arrow) is normal and extends posterior to mesenteric vessels. SMV = superior mesenteric vein, SMA = superior mesenteric artery, Py = pylorus, VB = vertebral body.

**TABLE 1: Sensitivity and Specificity of UGI Series, US, and Specific US Signs for the Diagnosis of Midgut Malrotation and Volvulus**

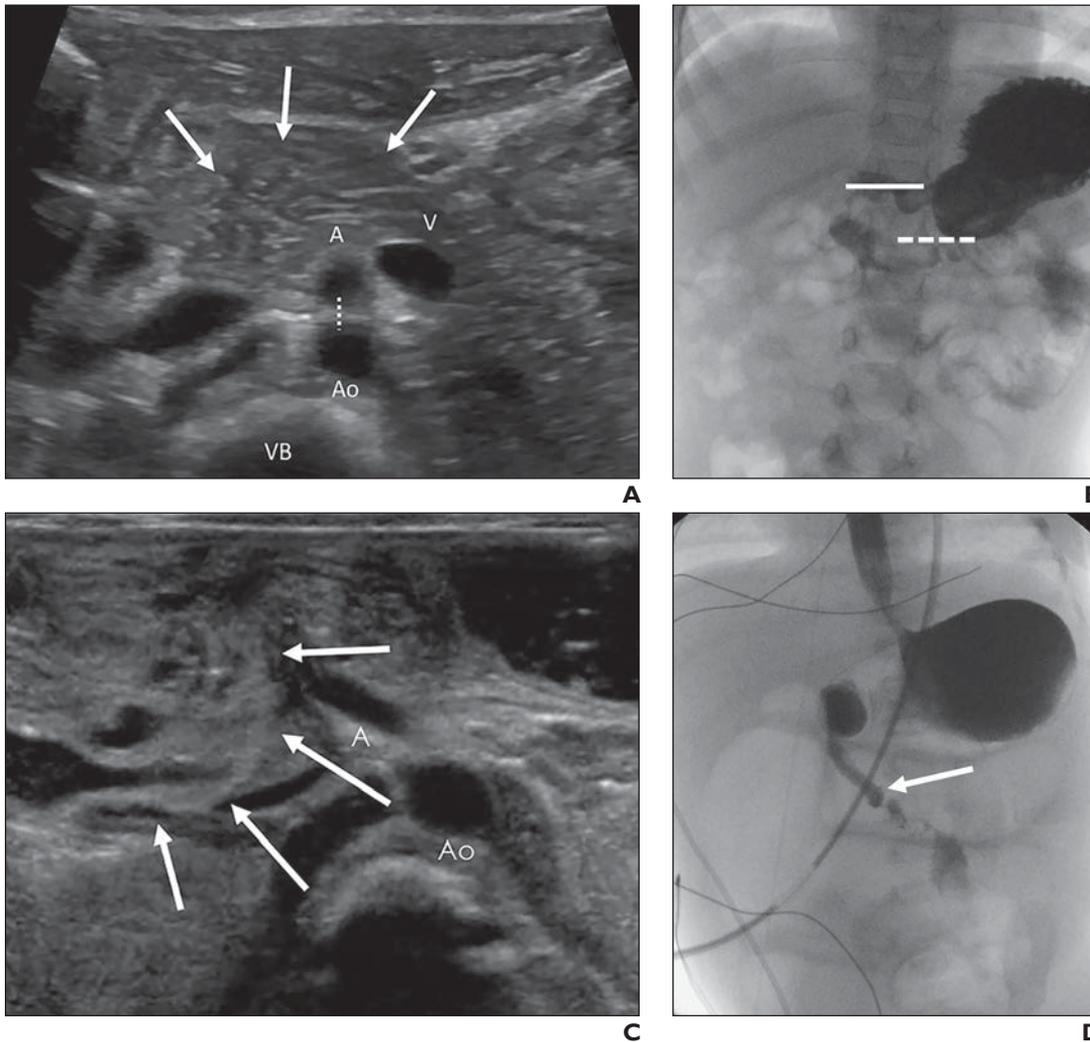
Imaging Test or Sign	Malrotation		Volvulus	
	Sensitivity (%)	Specificity (%)	Sensitivity (%)	Specificity (%)
<b>Index tests</b>				
UGI series	93–100 [44, 50, 51]	64–100 [15, 24, 26–28, 44]	54–79 [44, 51]	98 [44]
US	75–100 [10, 13, 15, 16, 24–28, 30, 58, 59]	54–100 [10, 16, 24, 25, 27, 28, 30, 58, 60]	83–100 [13–15, 23, 26, 28, 30]	91–100 [14, 15, 26, 30]
<b>US signs</b>				
Abnormal SMV and SMA relationship	30–100 [13, 15, 23, 24, 26, 32, 35, 59, 60]	93–100 [24, 26, 32, 35, 59]	NA	NA
Intraperitoneal course of D3 (anterior to SMA)	100 [19, 22, 24]	82–100 [19, 22, 24]	NA	NA
Hypoplastic uncinate process	NA	NA	NA	NA
Abnormal cecal position	NA	NA	NA	NA
Whirlpool sign	96 [24]	96 [24]	45–100 [13–16, 23, 26, 28, 30]	89–100 [14–16, 24, 26, 30]
SMA cutoff sign	NA	NA	100 [30]	100 [30]
Tapering or beaking of D3	NA	NA	89 [15]	91 [15]
Dilated proximal duodenum	NA	NA	100 [12]	100 [12]
Mesenteric edema	NA	NA	NA	NA
Ascites	NA	NA	33 [15]	82 [15]

Note—UGI = upper gastrointestinal, US = ultrasound, SMV = superior mesenteric vein, SMA = superior mesenteric artery, NA = not available, D3 = third portion of duodenum.

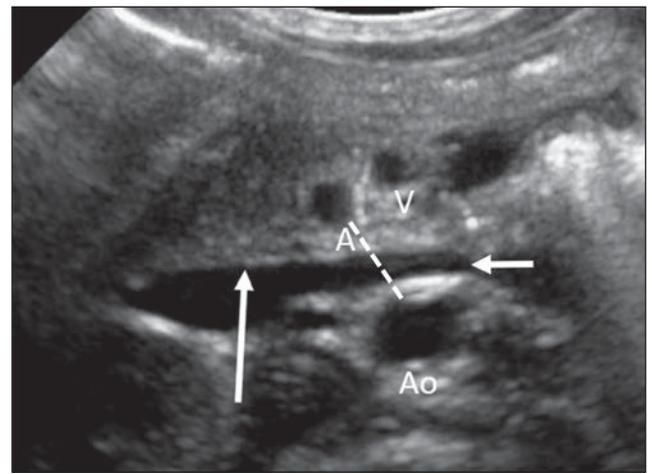
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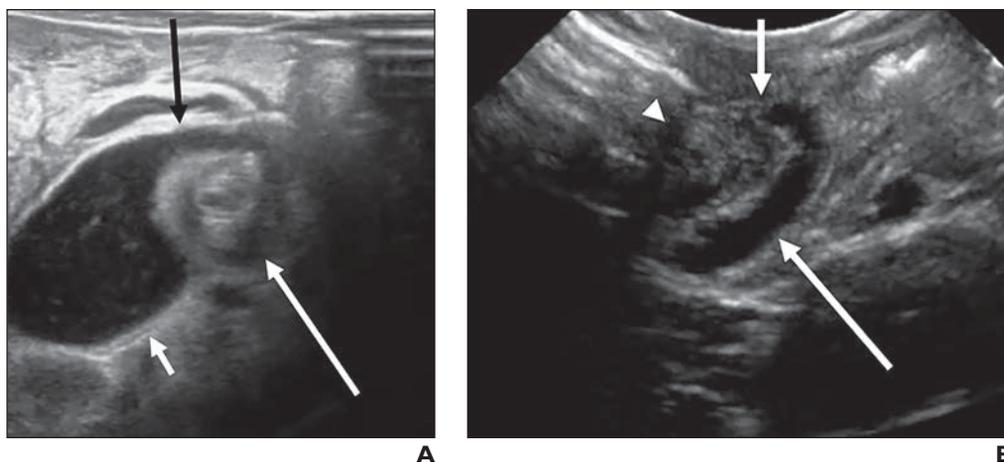
Volvulus is diagnosed on US by identification of a clockwise swirl (i.e., the whirlpool sign) of the SMV and its branches and of bowel loops around the SMA in the central abdomen when scanning in the transverse plane in a craniocaudal direction [13–15, 23, 26, 30, 37–39] (Figs. 3 and 4 and Video S2). The whirlpool

sign has a summary sensitivity of 92% and summary specificity of 99% [40]. Counterclockwise swirling of the jejunal branches of the SMV is a normal anatomic variant but may simulate a whirlpool sign and can be mistaken for volvulus [41, 42]. A key differentiating factor is the absence of twisting of bowel in as-



**Fig. 2**—Midgut malrotation in different patients. US = ultrasound, SMV = superior mesenteric vein, SMA = superior mesenteric artery, UGI = upper gastrointestinal. **A**, 2-year-old boy with fussiness. Transverse gray-scale US image at level of pancreas shows abnormal relationship between SMV and SMA-aorta axis: SMV (V) is located to left of axis formed between SMA (A) and aorta (Ao) (dotted line). Third portion of duodenum (arrows) is anterior to SMA. VB = vertebral body. **B**, Image from UGI series of same patient as in **A** shows duodenojejunal junction (dashed line) is slightly lower than pylorus and duodenal bulb (solid line). **C**, 1-day-old boy with emesis. Transverse gray-scale US image at epigastrium shows abnormal intraperitoneal course of third portion of duodenum, anterior to SMA (arrows), rather than retroperitoneal course between aorta (Ao) and SMA (A). **D**, Image from UGI series obtained 1 hour after **C** shows low duodenojejunal junction (arrow). **E**, 5-week-old girl with emesis. Transverse gray-scale US image at level of pancreas shows hypoplastic pancreatic uncinate process (long arrow) not extending posterior to mesenteric vessels. Video S2 is cine clip in this patient, which can be viewed in electronic supplement to this article at doi.org/10.2214/AJR.21.27242. A = SMA, V = SMV, Ao = aorta, short arrow = left renal vein, dashed line = SMA-aorta axis.





**Fig. 3**—Midgut volvulus in 5-day-old boy with bilious emesis. SMA = superior mesenteric artery, SMV = superior mesenteric vein.  
**A**, Transverse gray-scale ultrasound (US) image at epigastric region shows clockwise swirl (long white arrow) of SMV and bowel loops around SMA. Note also dilatation of second portion of duodenum (short arrow) proximal to point (black arrow) where it enters volvulus.  
**B**, Transverse gray-scale US image at periumbilical region shows SMV dilatation (long arrow) upstream to point (short arrow) where it enters volvulus (arrowhead).

sociation with the normal counterclockwise swirling of the jejunal veins.

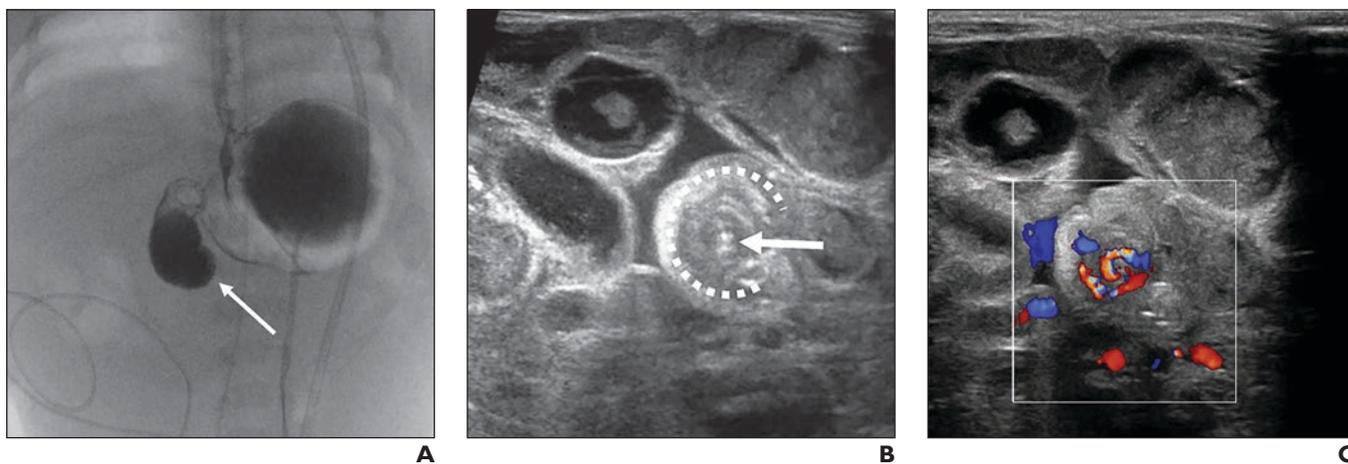
Ancillary US findings of volvulus include dilatation of D2 with to-and-fro flow of enteric contents, although this finding may be absent due to emesis or decompression by an enteric tube (Video S3, which can be viewed in the electronic supplement to this article at doi.org/10.2214/AJR.21.27242). Tapering of D3 as it becomes pulled into the volvulus, the SMA cutoff sign, dilatation of the SMV distal to the volvulus, mesenteric edema, and ascites have also been described [9–17, 19–31, 43] (Table 1, Figs. 3 and 4, and Video S2).

### Advantages and Disadvantages of Upper Gastrointestinal Series

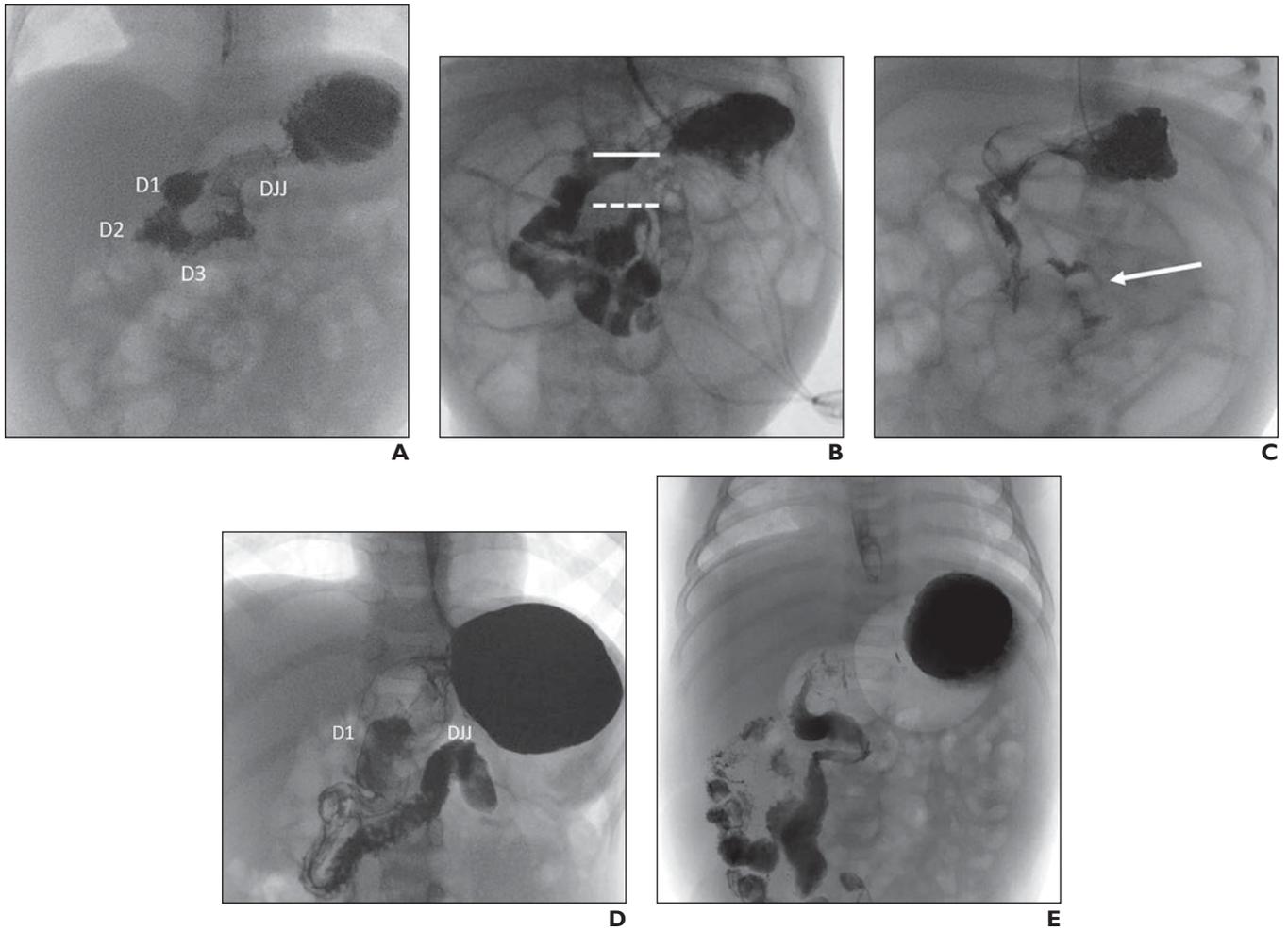
An advantage of the UGI series for the diagnosis of malrotation and volvulus is that it is familiar to referring physicians, surgeons, and radiologists. A systematic review and meta-analysis of UGI series for diagnosing malrotation with or without volvulus reported a high summary sensitivity and specificity of 91% and 94%, respectively [40]. However, few of the published studies on UGI series analyzed malrotation and volvulus separately or reported

the examination's true-negative rate. Another advantage of the UGI series is that, in cases that are equivocal for malrotation, a concurrent contrast enema or the conversion of the UGI series into a small-bowel follow-through series allows determination of the location of the cecum and thus an inference of the length of the mesenteric root (i.e., the distance between the duodenojejunal junction and cecum). However, in infants and in some older children, a lax mesocolon occasionally leads to a mobile cecum, limiting inference of the length of the mesenteric root.

Although the UGI series represents the traditional standard of care in diagnosing malrotation and volvulus, it has shortcomings. The UGI series is customarily not performed portably, limiting its accessibility for clinically unstable or critically ill patients who are unable to travel or to be monitored in the radiology department, as well as limiting the ability to perform the study in the absence of an on-site radiologist. Furthermore, the UGI series is operator-dependent and, even with meticulous attention to technique, may yield false-positive, false-negative, and equivocal results in the diagnosis of malrotation without or with volvulus. The false-positive and false-negative rates across individual published studies range



**Fig. 4**—Midgut volvulus in 2-day-old boy (33 weeks' gestation) with bilious emesis.  
**A**, Image from upper gastrointestinal series shows abrupt stop of contrast progression at second portion of duodenum (arrow), which is consistent with complete obstruction and suspicious for midgut volvulus.  
**B** and **C**, Gray-scale (**B**) and color Doppler (**C**) ultrasound images of mid abdomen show swirling (curve, **B**) around superior mesenteric artery (arrow, **B**), which is described as whirlpool sign on color Doppler and is consistent with diagnosis of volvulus.



**Fig. 5**—Upper gastrointestinal (UGI) series in different patients. DJJ = duodenojejunal junction.

- A**, 2-month-old boy with persistent emesis. Normal duodenal C sweep—that is, DJJ located at same level as pylorus and duodenal bulb (D1)—is noted. Second (D2) and third (D3) portions of duodenum are indicated.
- B**, 2-month-old girl with bilious emesis. Midgut malrotation is suggested due to DJJ located at level below that of pylorus (*solid line*) and duodenal bulb (*dashed line*). Laparotomy identified dense adhesions of bowel to anterior abdominal wall and normal midgut rotation, indicating false-positive result for malrotation on UGI.
- C**, 11-month-old boy with yellow emesis. Midgut volvulus was suggested due to subtle corkscrew appearance (*arrow*). Surgery confirmed malrotation but no volvulus, indicating false-positive result for volvulus on UGI series.
- D**, 2-year-old boy with emesis and abdominal pain. UGI series is normal; for example, radiograph from UGI series shows normal position of DJJ at level of pylorus and duodenal bulb (D1). Abdominopelvic CT (not shown) performed for appendicitis identified appendix and colon in left lower abdomen associated with abnormal superior mesenteric vein–superior mesenteric artery relationship, suggesting malrotation and indicating false-negative result for malrotation on UGI series.
- E**, 1-month-old boy with bilious emesis. Diagnosis based on UGI series was midgut malrotation without obstruction to contrast progression; midgut volvulus was not definitively diagnosed. Surgery showed malrotation with volvulus, indicating false-negative result for volvulus on UGI.

from 1% to 18% and from 1% to 8%, respectively [44–48]. For example, in the presence of an obstructed duodenum, a redundant duodenum, or a borderline low or medialized duodenojejunal junction, the UGI series may be falsely positive (Fig. 5). The diagnosis of malrotation and volvulus may also be difficult on UGI series given reliance on the projection of a 3D phenomenon (i.e., the spiraling of bowel around the mesenteric pedicle) on a two-plane image [3, 7, 8, 16, 40, 42, 44, 45, 47–55] (Fig. 5).

### Advantages and Disadvantages of Ultrasound

The portable nature of US avoids the need to transport a potentially unstable patient to perform the examination. Indeed, when performing the examination of patients in the neonatal

ICU (a common setting for evaluation of volvulus), it is expected that less time is needed for travel by a sonographer or radiologist to the ICU than would be needed for the mobilization and transport of the patient and support team from the ICU to the fluoroscopy suite. Moreover, US is being increasingly adopted for evaluation of necrotizing enterocolitis, and volvulus and hypertrophic pyloric stenosis may be assessed as an alternative diagnosis in the same imaging session.

Another advantage of US in the evaluation of malrotation and volvulus is that, assuming sonographer expertise in the scanning technique, the radiologist does not need to be on-site. This allows more timely performance and interpretation of studies at remote locations and a decrease in the radiologist’s call respon-

sibilities, an important consideration in the context of burnout among pediatric radiologists [56]. Finally, though US has been shown to cost less than an UGI series for evaluation of volvulus, it has comparable performance with a summary sensitivity and specificity of 94% and 100%, respectively [40, 57].

The advantages of US are tempered by concern for the potentially catastrophic consequences of a missed diagnosis of volvulus. The false-positive and false-negative rates of US based on a meta-analysis are 0% and 6%, respectively [40]. Nonetheless, false-negative diagnoses for both malrotation and volvulus may also occur for UGI examinations.

US has several shortcomings. Probe positioning off midline may alter the apparent relationship between the SMA and SMV and may result in false-negative or false-positive diagnoses of malrotation [23, 58–61]. This pitfall may be avoided by including an assessment of the position of the SMV relative to the SMA-aorta axis [62, 63]. As with any new technique, the use of US for this diagnosis has an associated learning curve; initially performing a follow-up UGI series after US—even when the diagnosis on US is not equivocal—will help develop confidence in the diagnosis by US [55, 57]. Over time, the need for performing both studies will decrease. Also, US may be nondiagnostic due to obscuration of anatomy by bowel gas artifact, as reported in 8–23% of patients in prior series, thereby possibly delaying surgical care [16, 27, 30, 59]. Being prepared to perform an UGI series immediately after nondiagnostic US mitigates this potential care delay [55]. An established institutional clinical practice guideline, developed in a collaborative multidisciplinary fashion, helps streamline the decision-making process and prevent delays in diagnosis and management.

Finally, although not an inherent disadvantage of the modality of US, not all institutions have pediatric sonographers available on a 24/7 basis, and not all patients with these conditions will initially present at a children's hospital. These concerns may be mitigated by educating sonographers and radiologists beyond those specializing in pediatric radiology to achieve broader expertise in the technique in the community setting.

### Should Radiologists Transition to Diagnosing Malrotation and Volvulus by Ultrasound?

Professional society guidelines now support the use of US for the diagnosis of both malrotation and volvulus [64]. Although US has excellent reported performance for the diagnosis of both conditions that compares favorably with that of the UGI series (Table 1), most of the relevant studies are retrospective, the studies often use the terms “malrotation” and “volvulus” interchangeably, and the specific evaluated US findings for malrotation and volvulus vary across the studies. Prospective studies are needed to compare the diagnostic performance of US and the UGI series, including in terms of the sensitivity and specificity of individual findings for malrotation (e.g., the SMA-SMV relationship, abnormal D3 course, cecal position, and uncinata process hypoplasia) and for volvulus (e.g., D2 dilatation, bowel and vessel swirling on gray-scale and color Doppler US, SMA cutoff sign, mesenteric edema, bowel dilatation, and postvolvulus SMV dilatation).

Prospective studies should also assess the optimal combination of findings; the role (if any) of administration of fluid, either orally or by enteric tube, when performing US for this diagnosis; and the role (if any) of obtaining an abdominal radiograph to use

the amount of bowel gas to guide the decision of whether to perform US. Future studies should also seek to determine the optimal imaging pathway for different clinical settings, such as for institutions with limited after-hours US availability versus for institutions with 24/7 US availability, as well as for nonurgent ambulatory settings versus for emergent settings (e.g., patients with bilious emesis). Of particular interest is definition of the scenario or scenarios in which an UGI series is needed before or after US.

For US to become the primary imaging modality for the evaluation of malrotation and volvulus, coordinated efforts will be needed to maximize the likelihood of achieving a timely and diagnostic examination. Clinical practices will need to train their sonographers and radiologists in the examination's performance and interpretation and are advised to implement a standardized imaging protocol (Appendix 1) and report template (Appendix 2).

### Consensus Statements

- The choice between UGI and US for the diagnosis of malrotation and volvulus will depend on the local expertise and available resources. Developing expertise in US for this indication is encouraged as US has comparable diagnostic performance to the UGI series but is radiation-free, is generally less costly than the UGI series, and can be performed more expeditiously than the UGI series given its portability.
- Radiology practices should implement standardized protocols and report templates for the US evaluation of malrotation and volvulus to guide sonographers and radiologists in assessing all pertinent findings and clearly communicating the results.
- Routine US examinations performed for suspected pyloric stenosis may include assessment of the duodenum and superior mesenteric vessels to assist sonographers and radiologists in gaining and maintaining proficiency in the evaluation of malrotation and volvulus.
- When having the available local expertise and resources (including patient transport team for US, fluoroscopy room for possible subsequent emergent UGI series, and pediatric surgery service on standby) to perform emergent US to evaluate for suspected midgut volvulus, a standardized algorithm is recommended. If the US examination is positive for volvulus, then the patient proceeds to surgical evaluation; in this situation, a confirmatory UGI series is not necessary and incurs potential treatment delay. If US is negative for volvulus and malrotation and does not provide an alternative diagnosis, then the patient's presentation guides the decision between expectant medical management versus further imaging with the next appropriate test, which may include an UGI series. If US is inconclusive or nondiagnostic, then an emergent UGI series is performed.

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(Appendixes begin on next page)

## APPENDIX 1: Ultrasound Protocol for Suspected Malrotation and Volvulus

1. Use linear high-frequency probe in the Musculoskeletal setting. If images are obscured by bowel gas, then use a microconvex probe in the Bowel setting.
2. Perform imaging along the midline in the transverse plane.
3. Obtain still images of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) at the level of the pancreas.
4. Obtain cine images (If cine images are obscured by bowel gas, attempt to move bowel gas by placing the patient in the right lateral decubitus position or by using a microconvex probe.) including the following:
  - Entire view of SMA and SMV (from main portal vein to jejunal branches or dome of the urinary bladder) both with and without color Doppler imaging and in dual-screen if possible.
  - Entire view of SMA and SMV without color Doppler imaging to assess for the whirlpool sign.
  - Pylorus and duodenal bulb.
  - Third segment of the duodenum must connect back to the duodenal bulb and pass posterior to the SMA.
  - Cecum with appendix in right lower quadrant.

## APPENDIX 2: Ultrasound Reporting Template for Suspected Malrotation and Volvulus

TECHNIQUE: Gray-scale ultrasound of the bowel performed in all four abdominal quadrants.

COMPARISON: [None]

### FINDINGS

SMV and SMA-aorta alignment at level of pancreas: [normal/abnormal/not assessable]

D2 dilatation: [absent/present/not assessable]

D3 course: [between aorta and SMA/anterior to SMA/not assessable]

Uncinate process of pancreas: [normal/hypoplastic/not assessable]

Cecal position: [normal/abnormal/not assessed]

Swirling of vessels and bowel: [no/clockwise/counterclockwise/not assessable]

SMA cutoff sign: [yes/no/not assessable]

Mesenteric edema: [yes/no/not assessable]

Bowel dilatation: [yes/no/not assessable]

SMV dilatation: [yes/no/not assessable]

Ascites/fluid collections: [none]

Other: [none]

### IMPRESSION

[Impression: 1/2/3/4/5]

1 = No findings of midgut malrotation = normal SMV and SMA-aorta alignment and D3 course between aorta and SMA

2 = Findings of midgut malrotation = one or more of the following findings: abnormal SMV and SMA-aorta alignment, D3 course anterior to SMA, abnormal cecal position, or hypoplastic uncinate process of pancreas

3 = Inconclusive or nondiagnostic for midgut malrotation and volvulus

4 = No findings of midgut volvulus = no swirling of vessels and bowel and no SMA cutoff sign

5 = Findings of midgut volvulus = one or more of the following findings: clockwise swirling of vessels or bowel, SMA cutoff sign, or SMV dilatation

Alternative diagnosis or additional diagnosis: [none]

[Communication: critical result/significant findings]

Note—SMA = superior mesenteric artery, SMV = superior mesenteric vein, D3 = third portion of duodenum, D2 = second portion of duodenum.