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NEONATAL IMAGING

Ultrasound for malrotation and volvulus — point

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Abstract

Upper gastrointestinal series is considered the gold standard imaging test to evaluate for malrotation and midgut volvulus. US has been explored for this purpose in recent years and has been shown to be a good alternative because of its relatively good accuracy, fast learning curve for sonographers and radiologists, as well as convenience for children, clinical staff and radiologists in the neonatal intensive care unit. In this article we describe our experience and review the evidence supporting the use of emergency US primarily to diagnose midgut volvulus as well as its use as a problem-solving tool for the diagnosis of malrotation. The examination technique, normal and abnormal findings are described, with emphasis on the whirlpool and superior mesenteric artery cutoff signs to diagnose midgut volvulus.

Keywords Children · Gastrointestinal tract · Infants · Malrotation · Midgut · Ultrasound · Upper gastrointestinal series · Volvulus

Introduction

Undiagnosed malrotation, or more precisely midgut volvulus, can lead to vascular compromise in the territory of the superior mesenteric artery pedicle and is well known for devastating consequences, including loss of extensive portions of small and large bowel, short gut syndrome and death [1].

Upper gastrointestinal (GI) series has been the preferred imaging modality to evaluate for malrotation and midgut volvulus for the last 50 years [2, 3]. Indeed, all pediatric radiologists are expected to know how to perform and interpret an upper GI series when malrotation or midgut volvulus is suspected [2, 3].

The interest in US to evaluate for malrotation began in 1987, with the first report of US to diagnose malrotation based on an abnormal relationship of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) in six cases [4]. A subsequent article published in 1992 reported the diagnosis of midgut volvulus based on the "whirlpool sign" [5]. A growing body of evidence supports a diagnostic role for US to evaluate children with suspected malrotation or midgut volvulus based on clinical presentation, recently summarized by a meta-analysis published by Nguyen et al. [6] in 2021. At our institution, US has been successfully implemented as the first-line imaging modality to evaluate for midgut volvulus since 2016, with buy-in from pediatric surgeons, neonatologists, radiologists and sonographers. In this article we briefly review the diagnostic accuracy of US and upper GI series for the diagnosis of malrotation with or without midgut volvulus and describe the US findings for midgut volvulus as well as malrotation without midgut volvulus, highlighting what we and others have perceived as advantages of US.

Accuracy of ultrasound and upper gastrointestinal series for the diagnosis of malrotation with or without midgut volvulus

The upper GI series is a well-established and reasonably accurate diagnostic test to detect malrotation with or without midgut volvulus, with a pooled sensitivity of 91% (95% confidence interval [CI]: 84–96%) based on recent meta-analysis data of 11 studies [7–17] that included 843 participants and were published between 2000 and 2018 [6]. The pooled specificity is also high, estimated at 94% (95% CI: 90–99%).

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However, considerable heterogeneity exists in the diagnostic accuracy among studies included in the meta-analysis, with sensitivities ranging from 40% in Zhou et al. [11] (2015) to 100% in Dekonenko et al. [17] (2019). Individual specificities ranged from 20% in Dilley et al. [8] to 100% in four other studies [7, 10, 14, 15]. In addition, the upper GI series has been reported as indeterminate for malrotation in 15–30% of young infants [18, 19], a troublesome fact because this is the population at highest risk for midgut volvulus.

The same meta-analysis [6] provided combined data of 17 studies [7, 11, 14–16, 20–31] reporting on the use of US for the diagnosis of small-bowel malrotation with or without midgut volvulus. These studies were published between 1987 and 2020 and included 2,257 participants. The pooled sensitivity was 94% (95% CI: 89–97%) and the pooled specificity was 100% (95% CI: 97–100%). The authors of the meta-analysis estimated that with a pooled prevalence of malrotation with or without volvulus of 17% (pre-test probability), the reported positive likelihood ratio of 317 would increase the post-test probability to 98%, whereas the negative likelihood ratio of 0.06 would decrease the post-test probability to 1%. Heterogeneity in diagnostic accuracy among studies was lower for US compared to upper GI series, with individual sensitivities ranging from 82% [16] to 100% [7, 11, 14, 21, 26, 28, 29] and specificities ranging from 55% [16] to 100% [7, 14, 15, 20, 21, 26, 28–31].

It can be concluded from the data that the upper GI series and US have similar sensitivity to diagnose malrotation with or without midgut volvulus (upper GI series, 91% with 95% CI: 84–96% vs. US, 94% with 95% CI: 89–97%; Fisher exact test P=0.5). US has a slightly higher specificity (upper GI series, 94% with 95% CI: 90–99% vs. US, 100% with 95% CI: 97–100%; P=0.04), implying a lower number of false-positive diagnoses for ultrasound [6]. The higher heterogeneity in individual sensitivities and specificities for the upper GI series suggests that there might be a higher inter-examiner variability in the performance or interpretation of the test compared to US. This might be explained by the not-infrequent technical or anatomical difficulties, the frequent lack of consensus for the upper GI technique even among examiners in the same institution, and the relatively frequent overlap between normal and abnormal findings (Fig. 1) [32].

Midgut volvulus

In an emergency situation, the most important question is whether there is midgut volvulus. It is not critical to answer all the questions such as the presence of uncomplicated malrotation or a duodenal web, for example, under pressing and sometimes suboptimal circumstances (e.g., middle of the night, trainee performing the exam, unstable or potentially unstable infant).

Previous studies have shown US to be accurate to answer this critical and pressing question, with a sensitivity for midgut volvulus ranging between 83% and 100% [5, 23, 31] and a specificity ranging between 92% and 100% [7, 23, 31]. US might be indeterminate in about 8% of cases [31].

Since the implementation of US as the initial imaging modality to evaluate for midgut volvulus at Phoenix Children's Hospital in 2016, we have not relied on the SMA–superior mesenteric vein (SMV) relationship to diagnose midgut volvulus because the SMV is often compressed and therefore not visible by US. Instead, because midgut volvulus occurs around the SMA, we simply follow the SMA from its origin down in the transverse plane looking for two signs: (1) the whirlpool sign and (2) the SMA cut-off sign (Fig. 2).

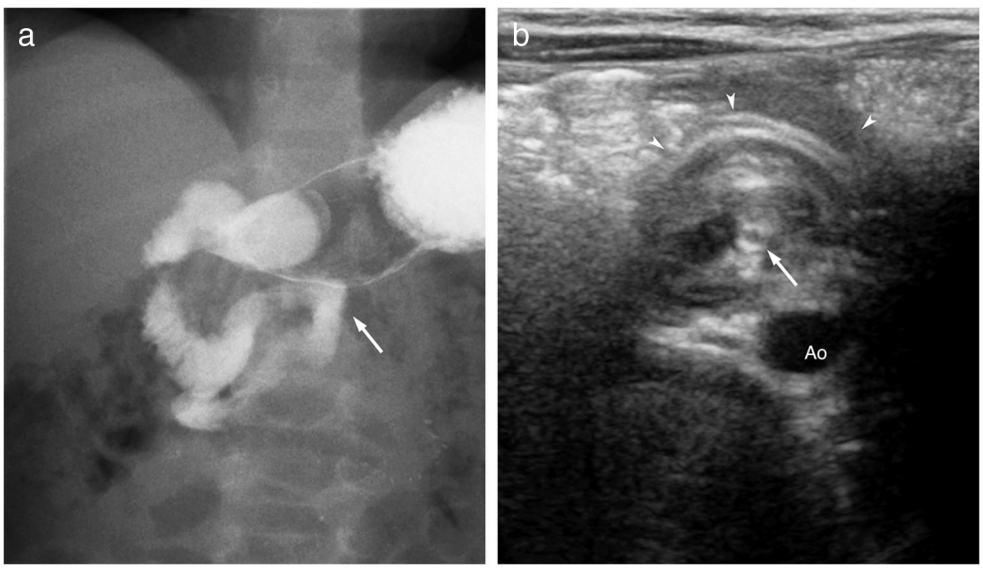


Fig. 1 Inconclusive upper gastrointestinal (GI) series in a 19-month-old girl. **a** Upper GI anteroposterior view shows the duodenojejunal junction (*arrow*) as slightly low and not to the left of the spine. This could be attributed to girl's rotation or malrotation. Nonurgent US was suggested. **b** US performed 5 h later. Transverse image of the epigastric region shows clockwise twist of the bowel (*arrowheads*) around the superior mesenteric artery (*arrow*), consistent with midgut volvulus. *Ao* aorta

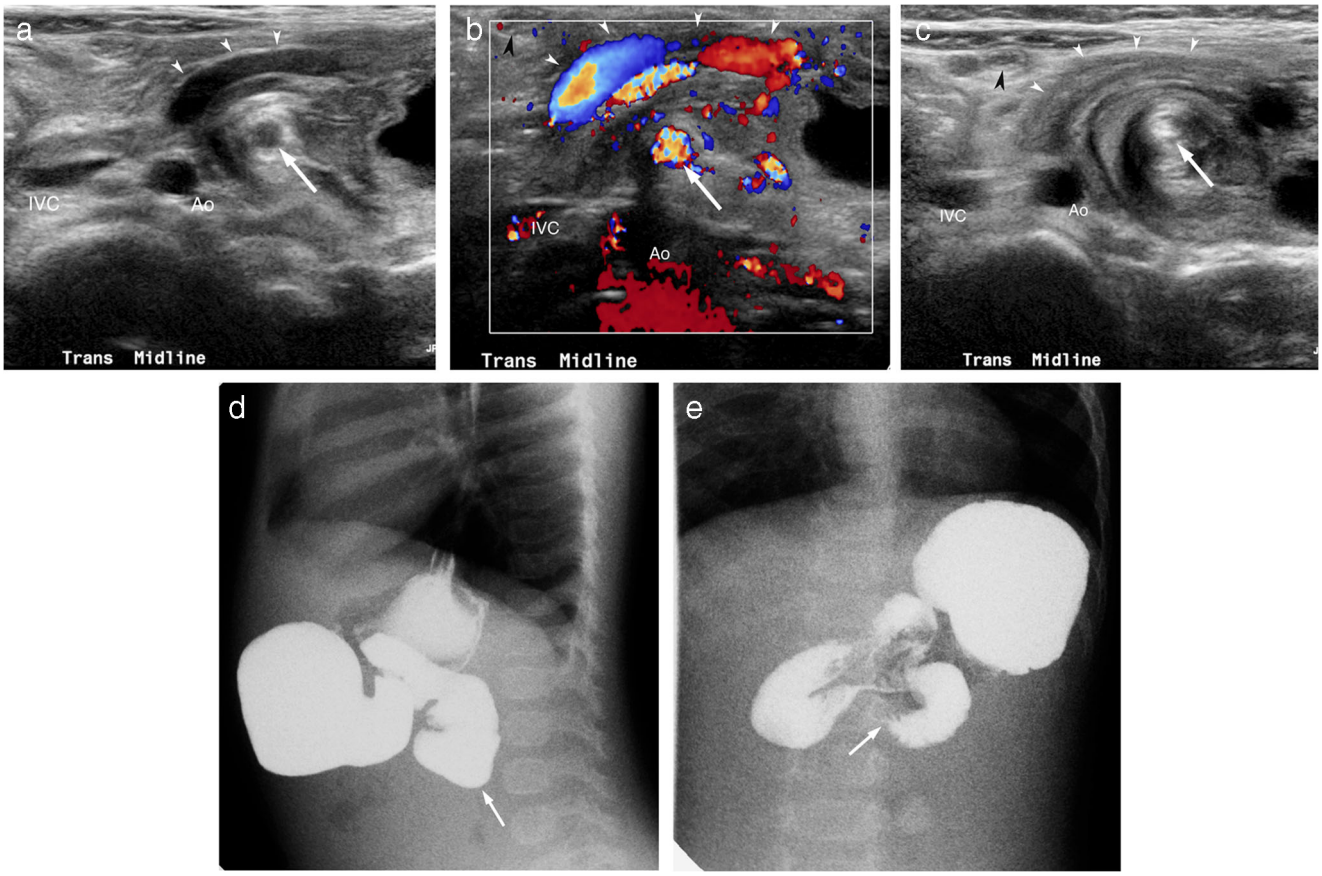


Fig. 2 Midgut volvulus in a 10-month-old girl. **a, b** Transverse US (**a**) and color Doppler US (**b**) images of the upper abdomen show the whirlpool sign and congested venous collaterals (*white arrowheads*) around the superior mesenteric artery (SMA) (*arrows*). *Black arrowhead* in (**b**) indicates abnormal position of the cecum/appendix. **c** SMA cut-off (*arrow*). Transverse US at another level shows clockwise twisting of the bowel (*white arrowheads*) and abnormal position of the cecum/appendix (*black arrowhead*). **d, e** Lateral (**d**) and anteroposterior (**e**) upper gastrointestinal series images 1 h after the US show dilated duodenum (*arrow* in **d**) and apparent beaking of the proximal jejunum (*arrow* in **e**)

Congested mesenteric venous tributaries along with SMA cut-off can be very helpful in cases with an abrupt twist (Fig. 3). We require sonographers to acquire and store cine clips because this makes it easier to appreciate the twisting of the bowel around the SMA. Cine clips also make it possible to review the studies

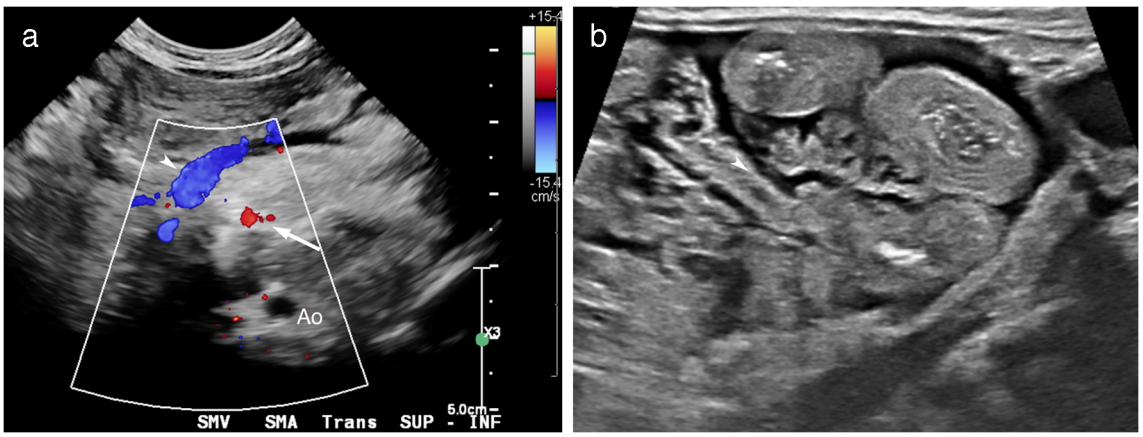


Fig. 3 Midgut volvulus around the superior mesenteric artery (SMA) without malrotation in a 15-day-old boy. **a** Transverse color Doppler US image shows a vanishing SMA (*arrow*) and congested venous collaterals (*arrowheads*). The twist was abrupt, better appreciated on cine clips (not shown). *Ao* aorta. **b** Transverse US image of left flank at a slightly higher level shows small bowel wall edema and ascites

performed by somebody else. Reviewing an upper GI series done by somebody else, even when cines from fluoroscopy are available, is not as easy in our experience.

Ultrasound has many obvious advantages over upper GI series, mainly US's lack of radiation and its portability and repeatability. US is particularly convenient for infants in the intensive care unit in situations where transporting them to the fluoroscopy suite might be difficult or impossible, aside from the potential difficulty in getting an adequate gastric emptying because of the sedation and drugs. Another obvious advantage of US over upper GI series is the avoidance of filling the stomach with contrast agent in a child who might require general anesthesia for an emergency surgery [33]. Although a nasogastric tube can be placed to aspirate the gastric contents, we feel that with US there is no added risk of aspiration or added time to suction out the contrast agent. Clinicians, in particular anesthesiologists, would prefer no additional filling of the stomach.

In our experience, US has also proved useful in children with congenital heart disease and known malrotation who might require repeated examinations to exclude midgut volvulus when they become symptomatic; in these children an elective Ladd procedure would preferentially be delayed until the cardiac condition is under control [34]. US is also very useful in children with duodenal obstruction in whom an upper GI series may or may not be able to exclude midgut volvulus without an understandable longer fluoroscopy time (Fig. 4). Another application is in the rare but critically important cases of midgut volvulus around the SMA without malrotation, in which upper GI series of the duodenojejunal junction would not be reasonably expected to diagnose (Fig. 3).

Ultrasound to diagnose midgut volvulus does not require extensive training [31, 33]. At institutions that have switched or are switching to US as a first-line imaging modality to diagnose midgut volvulus, evaluation of the SMA–SMV relationship, whirlpool sign, SMA cutoff and identification of a retroperitoneal third portion of the duodenum have been implemented as part of routine US protocols for evaluating other conditions such as pyloric stenosis and intussusception [31, 33]. This appears to contribute to the sonographers' experience and radiologist confidence in diagnosing midgut volvulus. As a result, the radiologist does not need to be in-house for the examination [31, 33]. US can be repeated if the images are inadequate or non-diagnostic. Radiation aside, repeating an upper GI series with residual contrast agent from a prior nondiagnostic examination can be challenging. In about 8% of the cases, the US examination might be inconclusive, mostly because of bowel gas [31]. In such cases, upper GI series should be obtained.

Malrotation

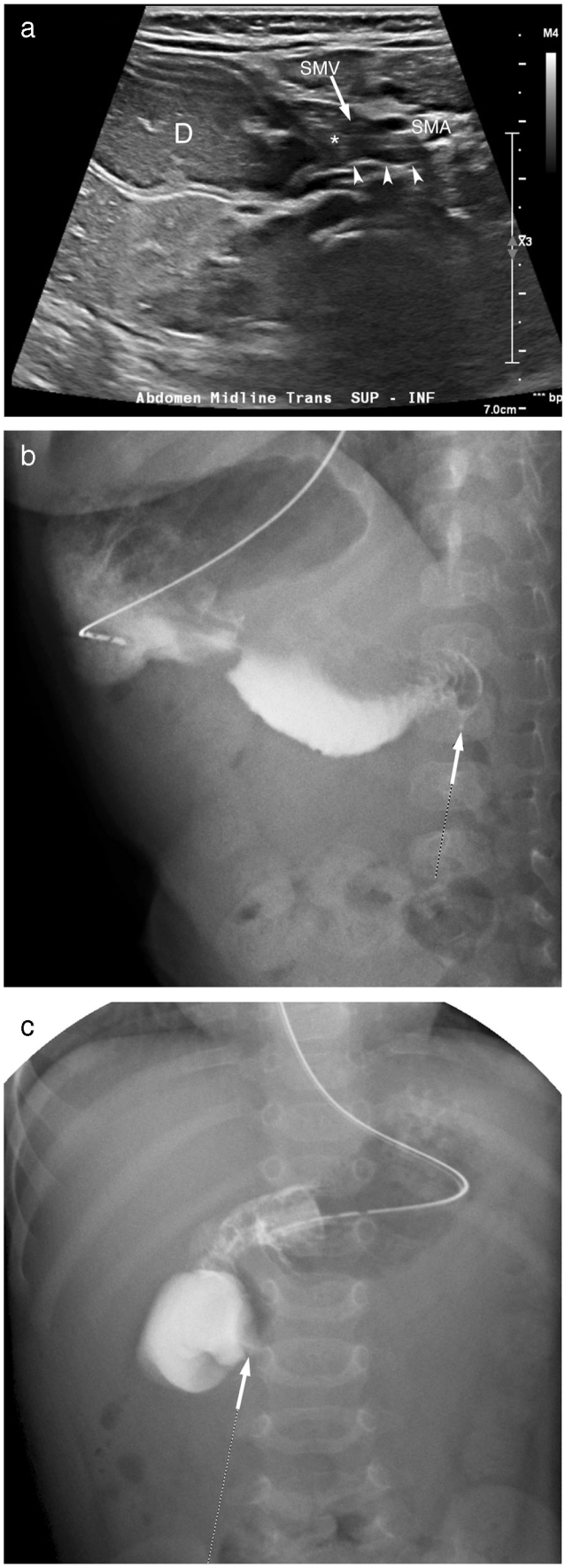
Upper GI series is routinely done first to assess for anatomical abnormalities to explain nonspecific symptoms such as abdominal pain, vomiting or failure to thrive. It is routinely carried out to the duodenojejunal junction to exclude malrotation despite the fact there is not solid evidence that malrotation without volvulus causes symptoms. Upper GI series is a good screening tool for multiple anatomical abnormalities including malrotation. However, in a busy practice, upper GI series is occasionally inconclusive despite meticulous technique in experienced hands, which sometimes requires longer fluoroscopy time. To deal with the not-uncommonly encountered inconclusive cases, some authors have suggested following the contrast agent to the cecum or even repeating the upper GI series [18]. US can be helpful in these inconclusive cases, increasing the degree of confidence through evaluation of the third portion of the duodenum, the SMA–SVA relationship, the pancreas uncinate process and the position of the cecum, as described in the following sections.

Retroperitoneal third portion of the duodenum

Based on embryological data, it has been suggested that a retroperitoneal third portion of the duodenum excludes malrotation [32]. This has been supported by some investigators with a sensitivity for malrotation of 97%, a specificity of 99% [35] and good correlation with upper GI series [36]. US has been shown to be a good tool in evaluating the position of the third portion of the duodenum in neonates [37]. The position of the third portion of the duodenum needs to be evaluated at the level of uncinate process of the pancreas. While a crossing bowel structure behind the SMA is not necessarily the third portion of the duodenum or necessarily retroperitoneal, aside from the third portion of the duodenum no normal bowel structures cross immediately contiguous to the uncinate process of the pancreas.

Superior mesenteric artery–superior mesenteric vein relationship

The mesenteric vessels relationship has been reported to have a US sensitivity ranging 67–98% [24, 38] and a specificity of 79% [24]. Based on the data available, a normal relationship does not exclude malrotation and an abnormal relationship does not always mean malrotation. However, the SMV is a relatively long structure and its relation to the SMA changes depending on the level of the scan. The reports don't specify at which level this relationship is best evaluated. The lack of cine



◀ **Fig. 4** Duodenal web in a 5-month-old boy. **a** A transverse US image at the level of the uncinate process of the pancreas (*asterisk*) shows normal superior mesenteric artery (*SMA*)–superior mesenteric vein (*SMV*) relationship (SMV is collapsed). The duodenum (*D*) is dilated and obstructed with a transition to a collapsed retroperitoneal third portion of the duodenum (*arrowheads*). No midgut volvulus is evident. **b, c** Lateral (**b**) and anteroposterior (**c**) upper gastrointestinal (GI) series images obtained the following morning show duodenal dilatation and beaking (*arrows*). Knowing the US findings, the contrast agent was not followed to the duodenojejunal junction

clips, early branching of the SMV and the angle of the scan can lead to false positives and false negatives. In addition, the clock position of the SMV in relation to the SMA is not always mentioned. This might explain the wide range of sensitivity for US in malrotation. Some investigators found the SMV to the right of the SMA, between 7 o'clock and 11 o'clock in 96% of individuals with normal rotation [36]. For consistency, we like to evaluate the SMA–SMV at the level of the uncinate process of the pancreas (Fig. 5).

Some reports looked at the combination of criteria to exclude malrotation, which has resulted in increased accuracy [39]. We have found that a US criteria combination of a retroperitoneal third portion of the duodenum and a normal SMA–SMV relationship increases the sensitivity of US for malrotation.

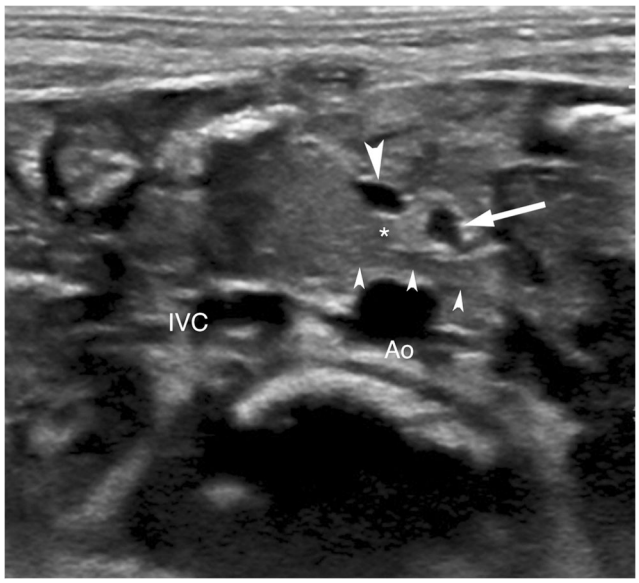


Fig. 5 Normal bowel rotation in a 5-day-old girl. Transverse US of the upper abdomen at the level of the uncinate process of the pancreas (*asterisk*) shows a normal relationship of the superior mesenteric artery (SMA; *arrow*) and superior mesenteric vein (SMV; *large arrowhead*), with the SMV to the right of the SMA. At this level, the normal retroperitoneal crossing of the third portion of the duodenum (*small arrowheads*) can also be appreciated. Note a normal uncinate process of the pancreas. *Ao* aorta, *IVC* inferior vena cava

The pancreatic uncinate process

The pancreatic uncinate process is the lower portion of the pancreatic head that lies dorsal to the superior mesenteric vessels. Because the development of the pancreas is intimately related to the rotation of the duodenojejunal loop in utero, abnormal rotation of the intestine might interrupt the normal rotation of the pancreatic primordia and result in an abnormal morphology of the uncinate process. Some authors have shown that the uncinate process is hypoplastic in malrotation [40]. While the accuracy of this observation has not been fully evaluated, we think that this is an additional finding that might add to the diagnostic confidence (Fig. 6).

The position of the cecum

While a cecum in the right lower quadrant is seen in 13–40% of patients with malrotation [1], it is well known that it this finding makes midgut volvulus very unlikely. It has been recommended to follow the contrast agent to the cecum in cases of an inconclusive upper GI series. Along with evaluating the third portion of the duodenum and the SMA–SMV relationship, US can be used in inconclusive upper GI exams to determine the location of the cecum by identifying the appendix. US is routinely used to diagnose appendicitis and therefore could be used to locate the cecum.

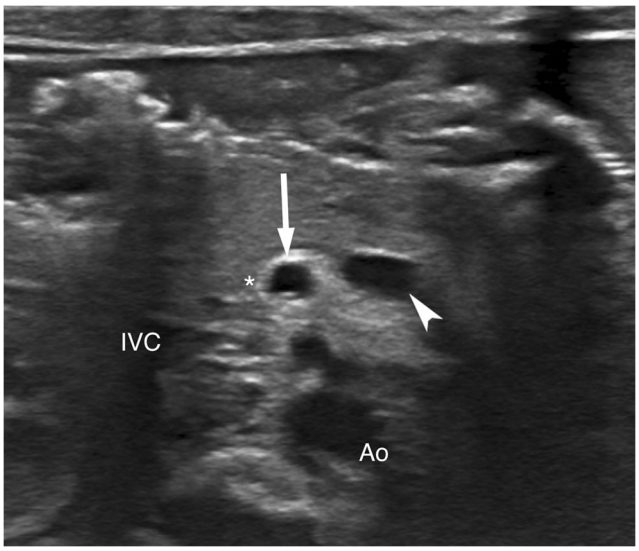


Fig. 6 Malrotation without midgut volvulus in a 20-day-old boy. Transverse US of the upper abdomen at the level of the uncinate process of the pancreas (*asterisk*) shows a reversed superior mesenteric artery (SMA)–superior mesenteric vein (SMV) relationship, with the SMV (*arrowhead*) to the left of the SMA (*arrow*). The uncinate process is hypoplastic. No retroperitoneal crossing of the third portion of the duodenum is identified. *Ao* aorta, *IVC* inferior vena cava

A note of caution when interpreting the literature

It should be noted that, when interpreting the available literature regarding the diagnostic accuracy of upper GI series or US to diagnose bowel malrotation and midgut volvulus, the two entities are sometimes lumped together as a single disorder [7, 11, 14, 20, 24–28, 30]. Aside from the fact that midgut volvulus is an emergency while malrotation is not, assessing these entities simultaneously can lead to difficulty in the interpretation of the results and conclusions of the study. Thus, we suggest that, moving forward, it would be more appropriate to evaluate the accuracy of diagnostic tests for each of these entities separately (i.e. malrotation with midgut volvulus and malrotation without midgut volvulus).

Conclusion

Ultrasound is a great tool to include or exclude midgut volvulus. Upper GI series should be reserved for rare cases in which US is equivocal or non-diagnostic. Upper GI series is routinely done to exclude multiple anatomical abnormalities. It is also accepted, although sometimes reluctantly, as the screening tool for malrotation in symptomatic children. In occasionally inconclusive cases, we have found that US can be used as a problem-solving tool or at least add some degree of confidence. For the diagnosis of malrotation, we like to think of upper GI series and US not as competing modalities but as complementary.

Declarations

Conflicts of interest None

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