

Received: 2023.11.02

Accepted: 2024.01.17

Available online: 2024.01.26

Published: 2024.03.14

Navigating Inconclusive Upper-Gastrointestinal Series in Infantile Bilious Vomiting: A Case Series on Intestinal Malrotation

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Manuscript Preparation E
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Financial support: None declared

Conflict of interest: None declared

Case series

Patients: Female, 1-day-old • Male, 4- month-old • Female, 3-day-old

Final Diagnosis: Intestinal malrotation

Symptoms: Bilious vomiting

Clinical Procedure: —

Specialty: Pediatrics and Neonatology • Radiology

Objective: Unusual clinical course


Background: Bilious vomiting in a child potentially portends the dire emergency of intestinal malrotation with volvulus, necessitating prompt surgical management, with differentials including small-bowel atresia, duodenal stenosis, annular pancreas, and intussusception. Although the upper-gastrointestinal series (UGI) is the diagnostic investigation of choice, up to 15% of the studies are inconclusive, thereby posing a diagnostic challenge.

Case Reports: We report a case series of 3 children referred for bilious vomiting, whose initial UGI was inconclusive and who were eventually confirmed to have intestinal malrotation at surgery. The first child was a female born at 37 weeks with antenatally diagnosed situs inversus and levocardia, who developed bilious vomiting on day 1 of life. The duodenojejunal flexure (DJ) could not be visualized on the UGI because of faint opacification on first pass of the contrast and subsequent overlap with the proximal jejunal loops. The second child was a male born at 36 weeks, presenting at age 4 months with bilious vomiting of 2 days duration. The third child was a female born at 29 weeks, presenting with bilious aspirates on day 3 of life. UGI for all 3 showed persistent hold-up of contrast at the proximal duodenum with no opacification of the distal duodenum or small bowel. Adjunctive techniques during the UGI and ultrasound examination helped achieve a preoperative diagnosis of malrotation in these children.

Conclusions: Application of diagnostic adjuncts to an inconclusive initial UGI may help elucidate a preoperative diagnosis of intestinal malrotation in infantile bilious vomiting.


Keywords: Fluoroscopy • Infant • Ultrasonography, Doppler • Volvulus of Midgut • Vomiting

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/943056>

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Background

An upper-gastrointestinal series (UGI) is usually the investigation of choice for patients who are clinically suspected to have intestinal malrotation. However, the imaging features in approximately 3-15% of UGI are inconclusive and may result in false-negative or false-positive inferences [1,2]. To increase diagnostic accuracy, techniques for optimal image acquisition and adjunctive imaging modalities may be utilized. We describe 3 cases in our institution in which adjunctive techniques and imaging modalities clarified an initially inconclusive UGI. There is currently no evidence-based decision-making algorithm on how to proceed when faced with an inconclusive UGI in an infant with bilious vomiting. To this end, we propose a diagnostic approach based on these illustrative cases and a literature review on the topic.

Case Reports

Patient 1

This was a female child born at 37 weeks with antenatally diagnosed situs inversus and levocardia, who developed frequent regurgitation and 1 episode of bilious vomiting on day 1 of life. She was otherwise alert and active with a non-distended and soft abdomen. Laboratory markers including blood gas on initial presentation did not show significant derangement. A chest and abdominal radiograph showed gastric bubble in the right hypochondrium (blue circle) in keeping with situs inversus, but no dilated bowel loops (Figure 1). Real-time UGI showed initial faint opacification and subsequent considerable overlap of the duodenal and proximal jejunal loops, with the duodenojejunal (DJ) flexure not elucidated (Figure 2).

The nasogastric tube was advanced into the duodenum and contrast was injected to selectively opacify the duodenum. This selective duodenography revealed the duodenal loop and DJ flexure on the left side of the spine, with the latter below the duodenal bulb level. No definite “corkscrew” appearance of the opacified small bowel loops typical for midgut volvulus was visualized. The opacified small-bowel loops were in the left side of the abdomen (Figure 3). Further images revealed the caecum in the epigastric region and colonic loops on the right side of the abdomen (Figure 4A). Together with a DJ flexure left of midline, which was seen on the preceding selective duodenography (Figure 4B), imaging features suggested a narrow mesenteric pedicle with attendant risk of midgut volvulus.

At laparotomy, the DJ flexure was to the left of the midline and partly abdominal in location. This abdominal duodenum had developed 2 hairpin turns with associated adhesions seen over the second and third parts of the duodenum, which were

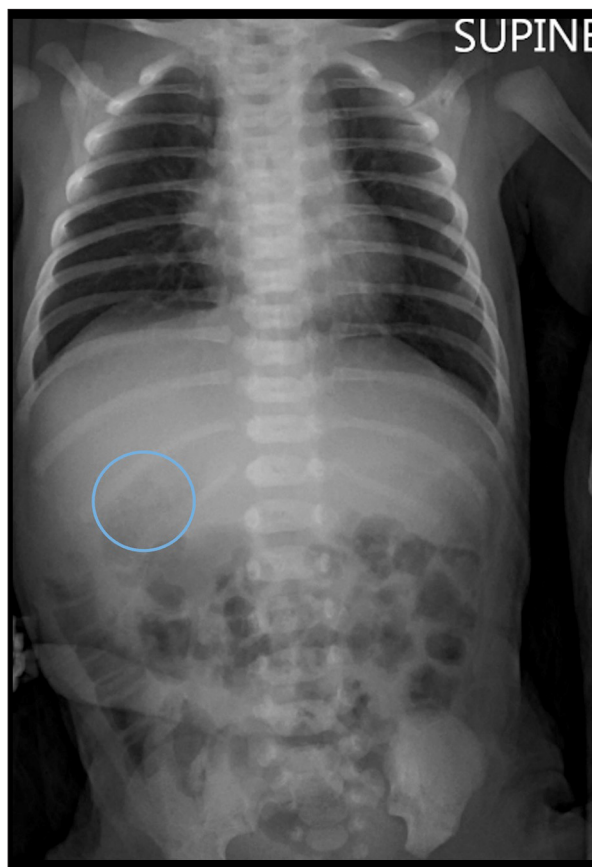


Figure 1. Chest and abdominal radiograph showing gastric bubble in the right hypochondrium (blue circle). There are no dilated bowel loops.

located to the left of the midline. The caecum was free-floating in the midline epigastrium with a narrow small-bowel mesenteric pedicle. The superior mesenteric vein (SMV) was on the right side of the superior mesenteric artery (SMA) (Figure 4C), representing reversal of SMV/SMA. There was no volvulus or bowel ischemia. A Ladd procedure was performed, with an uneventful post-operative recovery.

Patients 2 and 3

These were a male child born at 36 weeks, presenting at 4 months of age with bilious vomiting for 2 days duration, and a female child born at 29 weeks who presented with bilious aspirates on day 3 of life. The male child had no bowel output for 1 day duration, had high bilirubin levels, and dehydration with 11% weight loss. The patient’s blood gases did not show significant derangement. The female child was progressively unable to tolerate feeds with increasing bilious aspirates, and blood gases showed mild metabolic alkalosis.

Abdominal radiographs for both these patients similarly showed gas-distended stomachs with relative paucity of gas-distended

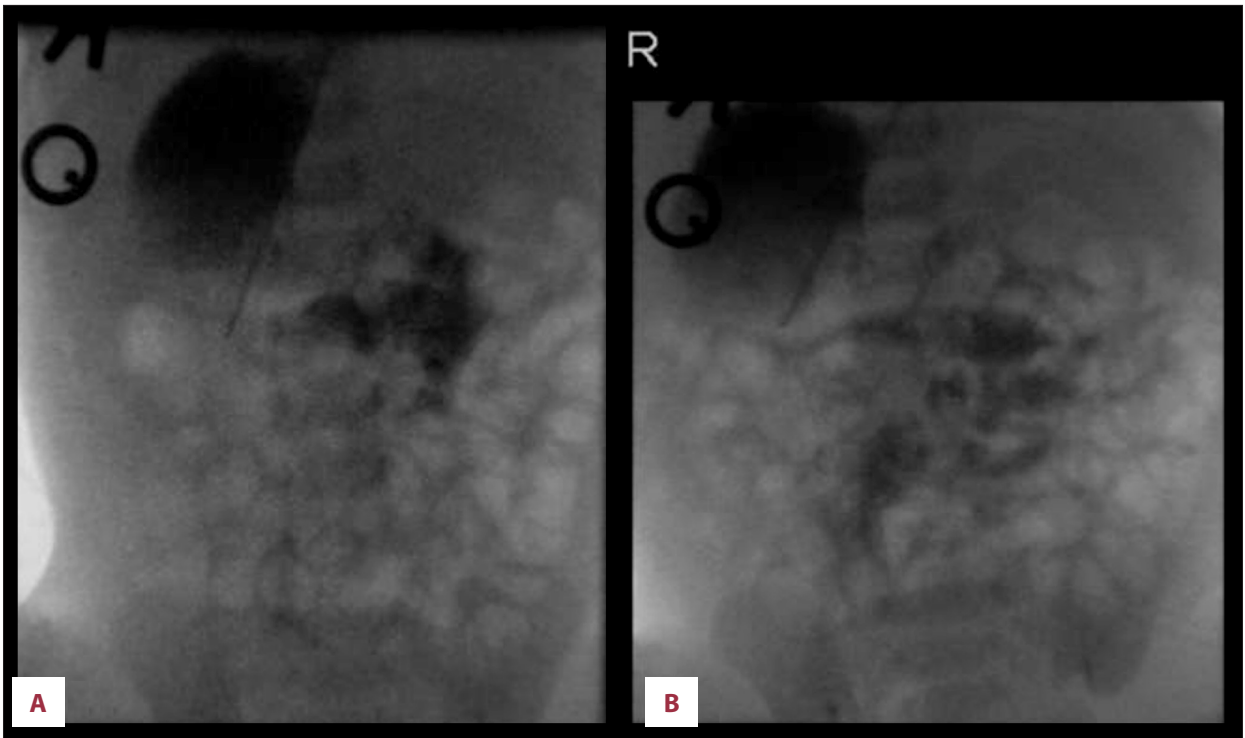


Figure 2. (A, B) Dextrogastria with position of duodenojejunal flexure not elucidated.

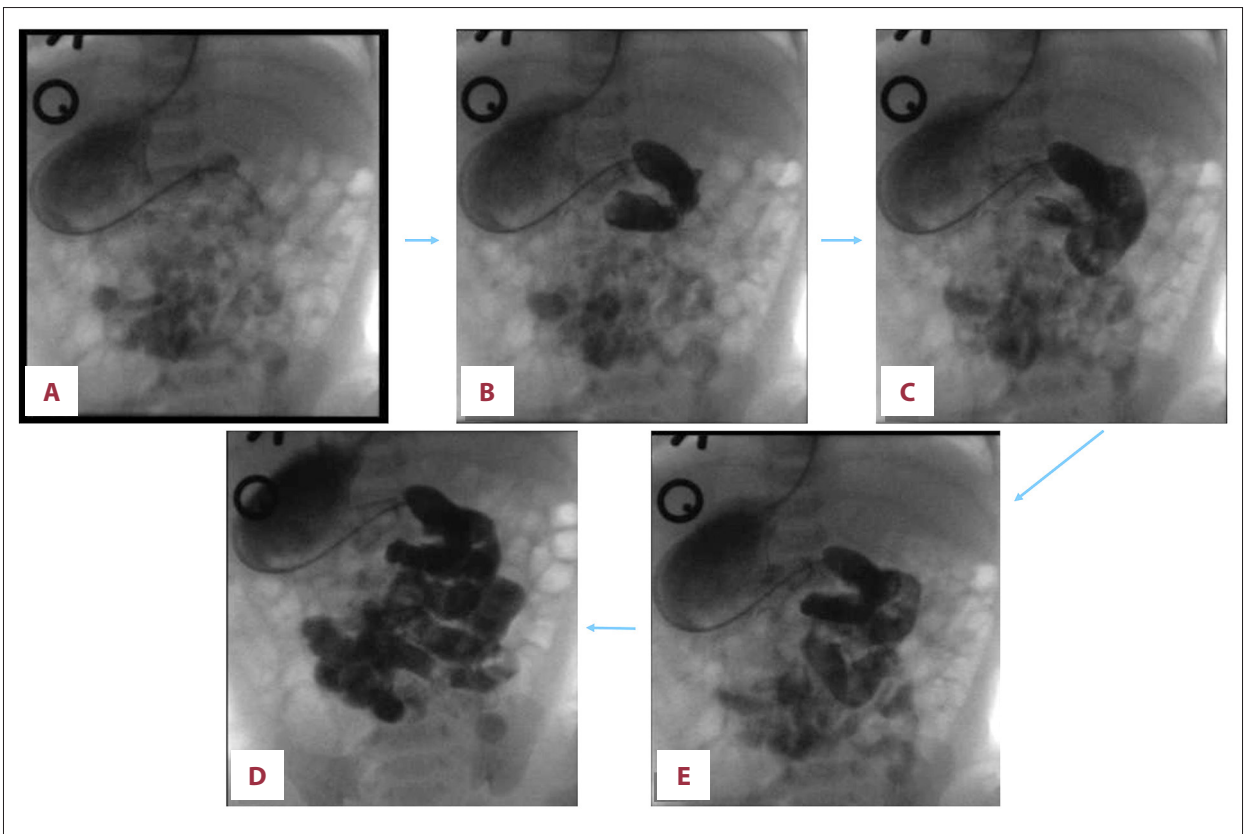


Figure 3. (A-E) Sequential grab images from selective duodenography revealed the duodenal loop and the duodenojejunal flexure on the left side of the spine, with the latter below the duodenal bulb level.

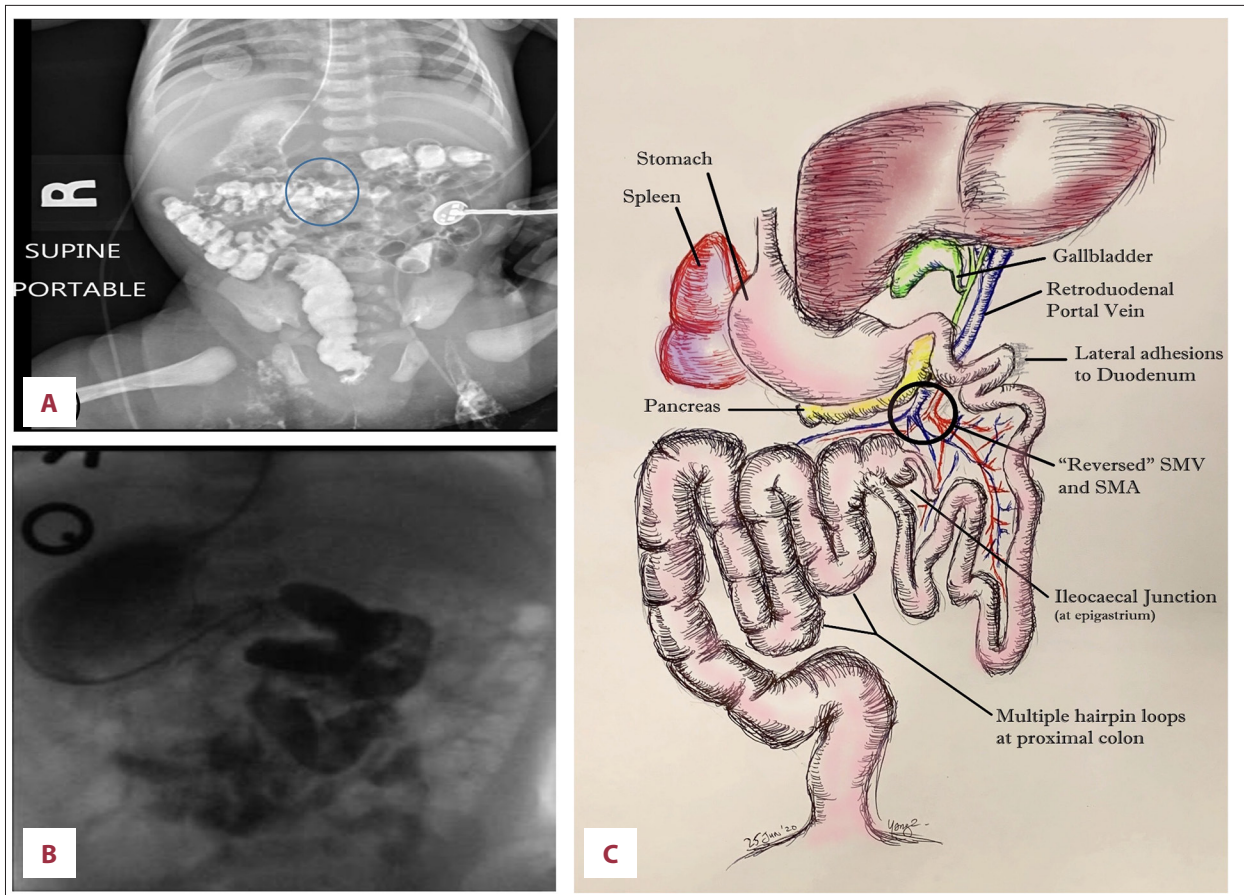


Figure 4. Additional follow-through radiograph (A) showed the cecum in the epigastric region (blue circle) and right-sided colonic loops. Together with a DJ flexure left of midline (B), imaging features suggested a narrow mesenteric pedicle with attendant risk of midgut volvulus. Pictorial diagram (C) shows congenital anatomic abnormalities found intra-operatively.

bowel loops elsewhere in the abdomen. UGI initially showed contrast flowing smoothly from the stomach into the proximal duodenum. Thereafter however, there was persistent hold-up of contrast at the proximal duodenum with no opacification of the distal duodenum or small bowel despite repeated repositioning and observation under intermittent fluoroscopy. An immediate ultrasound (US) showed an inverse SMA/SMV relationship, along with whirlpool sign of swirling of the mesenteric vessels (Figures 5, 6).

Intra-operatively, the male child was found to have intestinal malrotation with Ladd's band in the right lateral aspect, high-riding cecum next to the duodenum in the right hypochondrium, and narrowed root of mesentery. The female child had intestinal malrotation with caecum in the left hypochondrium and small bowel displaced to the right abdomen posterior to the caecum. The entire duodenum was on the right, with dense and broad Ladd's bands across the first and second part of the duodenum likely accounting for the obstruction. Both children had Ladd procedures and appendectomies with uneventful post-operative recovery.

The pertinent features of the cases presented are summarized in Table 1.

Discussion

Although most malrotation are diagnosed on an upper-gastrointestinal series, we illustrate the utility of known radiological adjuncts in our series of 3 cases, whose initial UGI studies were inconclusive. The UGI has been the imaging standard for diagnosing malrotation and associated volvulus in neonates and infants with bilious vomiting [2]. In acutely symptomatic but stable children, an urgent UGI is performed with an aim to expedite surgical treatment (Ladd's procedure) and avoid morbidity and mortality due to bowel ischemia caused by a twist in the mesentery.

The diagnosis of malrotation on UGI depends on careful delineation of the entire duodenum and the DJ on both anteroposterior and lateral projections [2]. This mandates real-time visualization of the passage of contrast through the duodenum

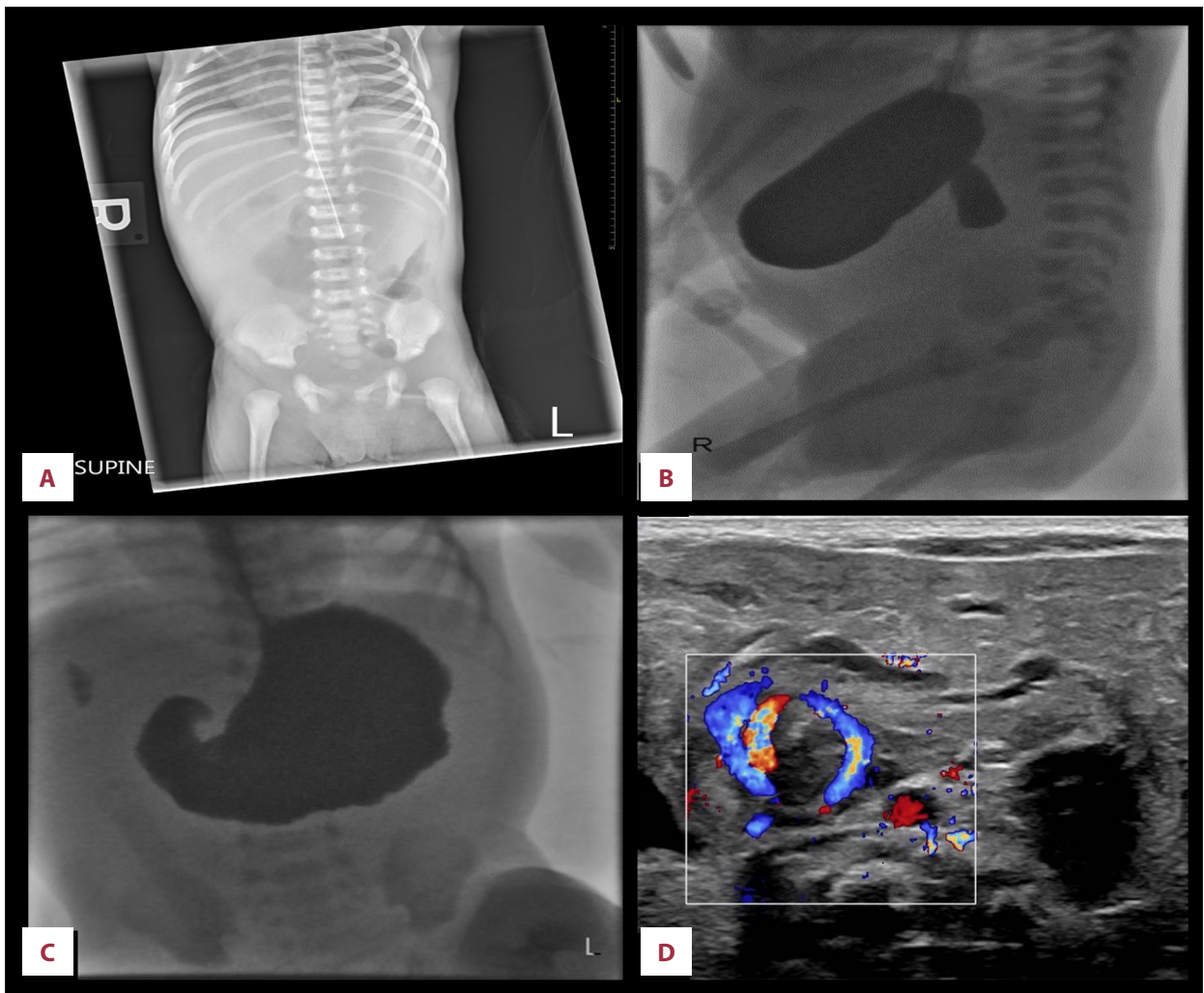


Figure 5. (A-D) Abdominal radiograph and upper-GI contrast study suggest a high-grade obstruction at the level of the proximal duodenum. US reveals reversal of superior mesenteric vessels.

on fluoroscopy to confirm the course of the duodenum and the position of the DJ. The diagnostic features of malrotation on an UGI are the flexure being on the right side of the midline and right-sided proximal jejunal loops [3]. However, these studies have a false-negative rate of 3% [4]. There are also false-positive rates of up to 15%, particularly if there is abnormal inferomedial positioning of the DJ flexure, which can occur in patients with enlarged spleens, lax ligaments, scoliosis, or gastric dilatation [3,4].

Contrast can be instilled via a nasogastric tube (NGT) for convenient and controlled instillation without having to depend on the child's swallowing, which can also expedite the procedure [5]. The passage of contrast into the duodenum thereafter depends on peristalsis aided by gravity with the child in the right-side-down position. This can result in trickling of contrast medium into the duodenum in advance of a bolus, inadequate distention of the duodenum with the first pass, and

then jejunal contrast medium obscuring the underlying duodenal loops, limiting the assessment [5]. In some cases, if contrast medium is not seen clearly flowing past the DJ flexure, it may not be clear if this is due to technical factors or due to true pathology, as in our first patient. While there is currently no consensus on how to proceed further after an inconclusive UGI, some adjunctive techniques and imaging modalities have been described to improve the diagnostic accuracy.

Selective Duodenography

The NGT can be purposefully advanced to lie in the proximal duodenum, allowing for performance of a "selective duodenogram" [5]. This allows the timing, pressure, and volume of the contrast medium bolus to be controlled, thereby expediting the procedure, possibly reducing radiation exposure, and resulting in distention of the duodenum during a contrast bolus. In the first child, selective duodenography allowed the

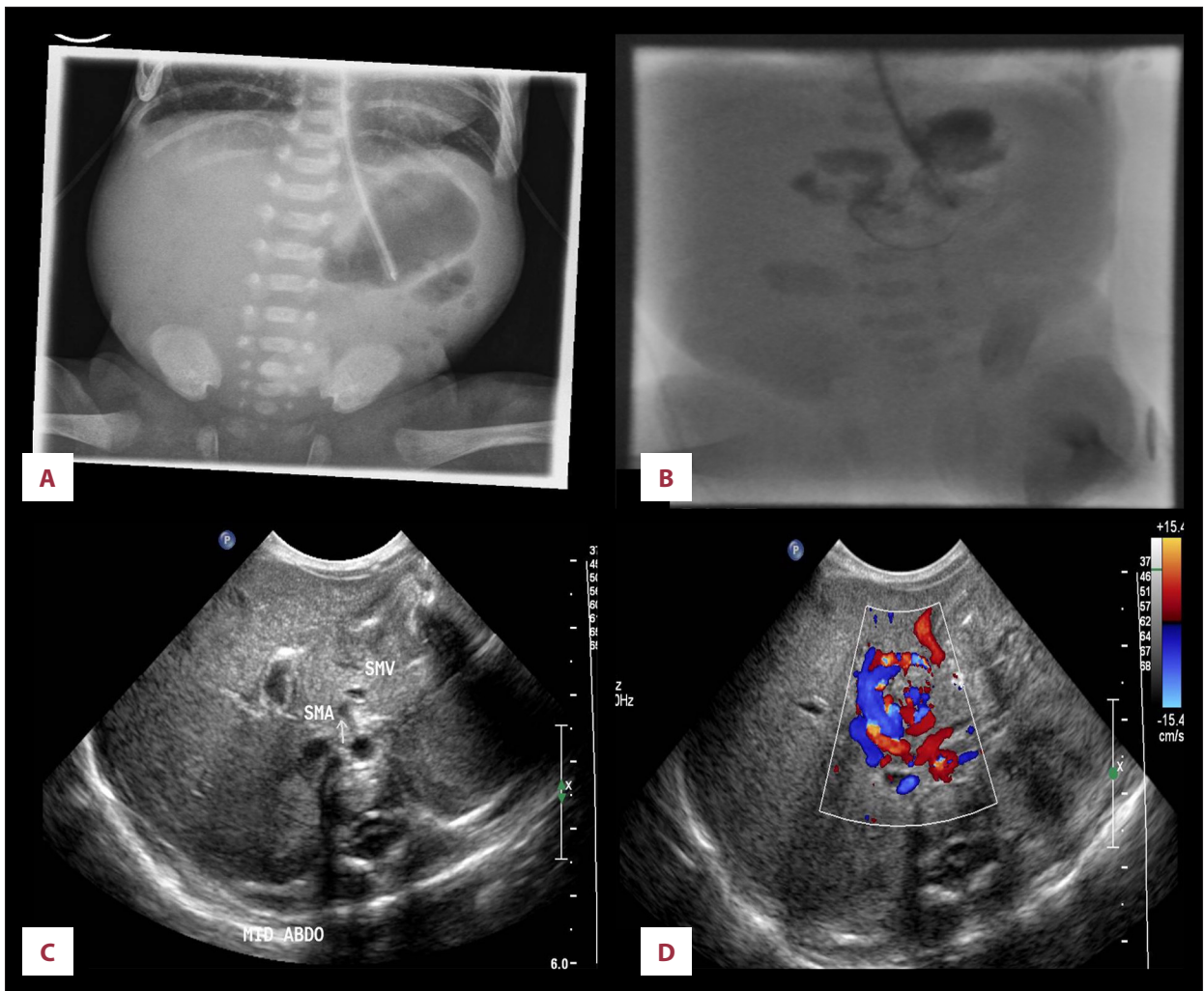


Figure 6. (A-D) Abdominal radiograph and upper-GI contrast study suggest high-grade obstruction at level of proximal duodenum. US findings suspicious for malrotation.

visualization of the DJ flexure and guided the distal passage of contrast thereafter. The technique involved pushing the indwelling NGT beyond the pylorus, and administering 2-3 ml of contrast bolus. A retrospective study by Andronikou et al in 101 patients showed that the selective duodenogram technique demonstrated the duodenum with 100% success, with significantly more frequent first-pass bolus visualization and duodenal distention than traditional studies [5].

Manual Epigastric Compression

In malrotation, the position of the DJ junction may not always be grossly abnormal, such that UGI can mimic a normal appearance [6]. When there is uncertainty, epigastric compression can be used to assess the DJ junction mobility. Usually, children aged less than 4 years old have lax bowel ligaments, which enables greater mobility of the DJ junction. In contrast, children with malrotation do not have mobile DJ junctions [2].

Manual epigastric compression involves applying a compression pressure no more than that applied for abdominal palpation during physical examination. In normal infants with a mobile duodenum, the DJ junction should return to a normal position after compression is released. In contrast, in patients with malrotation, the DJ junction remains in an abnormal position even after release of compression [6].

Ultrasound (US)

US has emerged as a complementary imaging modality to assess for malrotation. The intestinal whirlpool sign, along with the reversed SMA and SMV positions, have been shown to be highly accurate imaging findings for malrotation and volvulus [7]. As illustrated in our second and third patients, the inverse SMA/SMV relationship on US prompted urgent surgical intervention for intestinal malrotation. A retrospective study by Binu et al recommends the use of water as a contrast medium

Table 1. Summary table highlighting key information for each case.

Patients	Diagnostic challenges	Outcomes
<ul style="list-style-type: none"> Female child with bilious vomiting on day 1 of life 	<ul style="list-style-type: none"> UGI showed initial faint opacification and subsequent considerable overlap of the duodenal and proximal jejunal loops, DJ flexure not elucidated 	<ul style="list-style-type: none"> Selective duodenography revealed the duodenal loop and DJ flexure on the left side of the spine, with the latter below the duodenal bulb level. Further images revealed the cecum in the epigastric region and colonic loops on the right side of the abdomen. Imaging features suggested a narrow mesenteric pedicle with attendant risk of midgut volvulus A Ladd procedure was performed, with uneventful post-operative recovery
<ul style="list-style-type: none"> Male child with bilious vomiting at 4 months of age Female child with bilious aspirates on day 3 of life 	<ul style="list-style-type: none"> UGI showed persistent hold-up of contrast at the proximal duodenum with no opacification of the distal duodenum or small bowel despite repeated repositioning and observation under intermittent fluoroscopy 	<ul style="list-style-type: none"> US showed an inverse SMA/SMV relationship, along with whirlpool sign of swirling of the mesenteric vessels Both children had Ladd procedures and appendicectomies with uneventful post-operative recovery

under US guidance [3]. A fluid bolus is instilled via an indwelling NGT. After the pylorus is located, the passage of the fluid bolus is traced into the first part of the duodenum (D1). The transducer is then moved downwards to locate D2 as the fluid bolus progresses through the duodenum. Normally, the third part of the duodenum (D3) would course medially while traversing between the superior mesenteric vessels and the aorta. As the bolus is followed into D3, malrotation can be excluded by visualizing the normal retroperitoneal transverse course of D3. This would represent normal embryological development, in contrast to an intraperitoneal D3, which would be suspicious for malrotation. Finally, the transducer is moved upwards towards the SMA origin to follow the bolus into the fourth part of the duodenum (D4). Visualizing D4 with superior extent up to that of the SMA origin would demonstrate that the DJ flexure is in a normal position. In their study of 539 patients, all the patients who had US findings of malrotation had confirmatory intra-operative findings.

An expert panel narrative review by Nyugen et al recommends for a choice to be made between upper-GI contrast study and US depending on the local expertise and available resources [8]. The use of US for the evaluation of midgut rotation and volvulus was found to have comparable performance to that of UGI in terms of sensitivity and specificity, with additional benefits of being radiation-free, less costly, and possibility of being performed at bedside given its portability. They recommend a standardized algorithm when there are available local expertise and resources to perform emergent US. This involves performing US as the first-line investigation. If the US examination is positive for volvulus, then the patient proceeds

to surgical evaluation without the need of a confirmatory UGI, which can incur potential treatment delay. If the US is negative for volvulus and malrotation, and does not provide an alternative diagnosis, then the decision between expectant medical management versus further imaging would depend on the patient's presentation. If US is inconclusive or nondiagnostic, then an emergent UGI is performed.

A prospective study by Zhang et al showed that US could detect malrotation with higher accuracy than UGI, with the whirlpool sign indicating volvulus with a high accuracy of 92.3%. Furthermore, US is helpful in the assessing for the differential diagnoses of bilious vomiting, including annular pancreas and duodenal atresia [9]. Nonetheless, UGI, being the traditional preferred examination for the diagnosis of malrotation, remains a useful tool, and may still be considered first-line, particularly in centers where pediatric US may not be available [10].

In our institution, local experience is as yet unprepared for US to replace UGI as the first-line investigation for infantile bilious vomiting, or even fulfill the role of concurrent first-line investigation together with UGI, as routine practice. However, with the literature showing the merits of US, we anticipate US featuring even more prominently in our diagnostic algorithm of malrotation in the future, with further dedicated training and experience.

Delayed Radiograph or Contrast Enema

In equivocal cases of suspected malrotation without clinical features of volvulus, further follow-through images may be

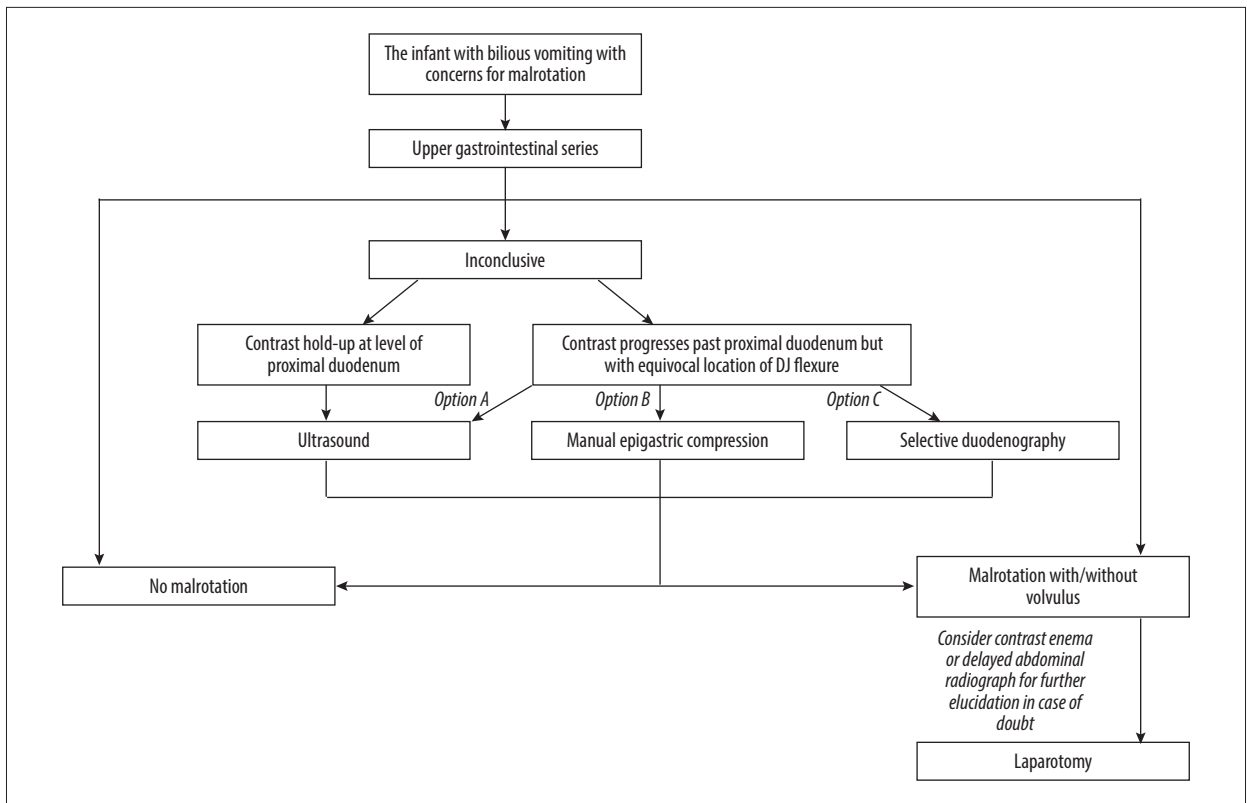


Figure 7. Proposed decision-making approach on how to proceed when encountering an inconclusive UGI in an infant with bilious vomiting.

helpful to delineate cecal position. Although normal cecal position may be observed in children with malrotation, an abnormal cecal position is seen in up to 80% of children with malrotation [2]. A contrast enema study may also be performed to expedite the identification of the position of the caecum and thus assess the length of mesenteric fixation [8].

CT/MRI

Malrotation can also be assessed on CT and MRI by the location of the small and large bowel, location of cecum, the presence or the absence of retroperitoneal duodenum, and the SMA-SMV relationship. CT and MRI studies can also help differentiate between malrotation and non-rotation. In non-rotation, the entire small bowel is located in the right abdomen and the colon is located in the left abdomen. CT and MRI have limited utility due to radiation and streak artefact from pre-existing GI contrast, and long duration of image acquisition, respectively [11]. Currently, CT and MRI are not considered in the routine evaluation of malrotation. They may however be useful in centers where pediatric UGI or US expertise is not available, especially given that a time-sensitive diagnosis is required in the setting of intestinal malrotation. CT and MRI can also be helpful for complex cases suspected to have alternative diagnoses [11,12].

In our institution, UGI remains the initial investigation of choice in the workup of children with bilious vomiting and clinical concerns for malrotation. The clinicians are in attendance for real-time feedback and decision-making during the UGI. Real-time visualization reveals gastric emptying/transit time and bowel peristalsis, together with potential point(s) of obstruction.

Proposed Decision-making Approach

We share 2 possible scenarios resulting in inconclusive UGI (Figure 7). In the first scenario with contrast hold-up up to the level of the proximal duodenum, we opine that persisting with further fluoroscopic techniques is of limited value with unwarranted radiation penalty to the patient, so we recommend early US evaluation in this scenario. We do not advocate use of selective duodenography in this instance, in view of potential safety concerns of advancing the NGT without knowledge of the cause of the contrast hold-up in the proximal duodenum. The non-opacification of the DJ flexure in this situation would also preclude the usefulness of manual epigastric compression. In the second scenario with contrast past the proximal duodenum but with equivocal DJ flexure position, manual epigastric compression and selective duodenography or US may clarify the diagnosis.

The cecum can be visualized either by performing additional follow-through radiographs during the UGI or an immediate contrast enema to rule out malrotation in an otherwise stable child [13].

Cross-sectional imaging modalities such as CT or MRI are not in routine use in the acute setting of suspected malrotation and volvulus in our institution.

Conclusions

Adjunct measures during the initial upper-gastrointestinal series or an expedited, targeted ultrasound in addition to UGI may clarify the preoperative diagnosis of intestinal malrotation in an infant who presents with bilious vomiting. The proposed

decision-making approach arises from our institution's experience and literature review, and therein lies the limitation of this study. Large-scale multicenter studies may reveal the efficacy of an integrated approach that routinely incorporates these adjuncts into the diagnostic algorithm of malrotation.

Department and Institution Where Work Was Done

Department of Diagnostic Imaging, National University Hospital, Singapore.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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