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ORIGINAL ARTICLE

Heterotaxy syndromes and abnormal bowel rotation

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Abstract
Background Bowel rotation abnormalities in heterotaxy are common. As more children survive cardiac surgery, the management of gastrointestinal abnormalities has become controversial.
Objective To evaluate imaging of malrotation in heterotaxy with surgical correlation and provide an algorithm for management.
Materials and methods Imaging reports of heterotaxic children with upper gastrointestinal (UGI) and/or small bowel follow-through (SBFT) were reviewed. Subsequently, fluoroscopic images were re-reviewed in conjunction with CT/MR studies. The original reports and re-reviewed images were compared and correlated with surgical findings.
Results Nineteen of 34 children with heterotaxy underwent UGI, 13/19 also had SBFT. In 15/19 reports, bowel rotation was called abnormal: 11 malrotation, 4 non-rotation, no cases of volvulus. Re-review, including CT (10/19) and MR (2/19), designated 17/19 (90%) as abnormal, 10 malrotation (abnormal bowel arrangement, narrow or uncertain length of mesentery) and 7 non-rotation (small bowel and colon on opposite sides *plus* low cecum with probable broad mesentery). The most useful CT/MR findings were absence of retroperitoneal duodenum in most abnormal cases and location of bowel, especially cecum. Abnormal orientation of mesenteric vessels suggested malrotation but was not universal. Nine children had elective bowel surgery; non-rotation was found in 4/9 and malrotation was found in 5/9, with discrepancies (non-rotation at surgery, malrotation on imaging) with 4 original interpretations and 1 re-review.
Conclusion We recommend routine, early UGI and SBFT studies once other, urgent clinical concerns have been stabilized, with elective laparoscopic surgery in abnormal or equivocal cases. Cross-sectional imaging, usually obtained for other reasons, can contribute diagnostically. Attempting to assess mesenteric width is important in differentiating non-rotation from malrotation and more accurately identifies appropriate surgical candidates.

Keywords Heterotaxy · Malrotation · Non-rotation · Children · Computed tomography · Magnetic resonance imaging · Upper gastrointestinal series

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Introduction

During fetal development, the small bowel undergoes a complex process of herniation, rotation (360°), reduction and fixation [1–3], resulting in the placement of the duodenum in a retroperitoneal position behind the superior mesenteric artery (SMA) [3–5], with subsequent duodenojejunal junction (DJJ) fixation to the retroperitoneum in the left upper quadrant (ligament of Treitz). There is separate, independent cecocolic rotation, which positions the cecum on the right with final fixation of the cecum to the retroperitoneum in the right lower abdomen, usually by the first few months of postnatal life [1, 3]. A broad-based SMA blood supply to the small intestine results from these widely spaced retroperitoneal fixation points [2, 3]. Failure to complete the process of normal bowel rotation (malrotation) results in both the DJJ and cecum

remaining in the mid upper abdomen producing a narrow mesenteric base and resultant risk of mesenteric vascular insufficiency and/or infarction following small bowel volvulus around the SMA [2, 6]. Ladd bands represent fibrous bands that occur variably between the cecum and retroperitoneum or between the abnormally closely apposed mesenteric leaves in the setting of malrotated midgut structures. Ladd bands are found and divided during surgery for malrotation to prevent their potential for obstructing the proximal duodenum or maintaining the mesenteric leaves in close apposition and, thereby, predisposing to volvulus [1, 3].

Non-rotation may also occur; this is a misnomer since the process is actually arrested after the first 90° of small bowel rotation [2, 3]. In this situation, the duodenum does not take a complete retroperitoneal course, and the DJJ is usually malpositioned to the right of the spine. The entire small bowel is located on one side of the abdomen (usually right) and all or most of the large bowel is on the other (usually left) [2, 3]. If the cecum is fixed in the lower abdomen, there is usually a sufficiently broad mesenteric base to prevent small bowel volvulus. Between classically defined malrotation and non-rotation, there are many partial forms of incomplete rotation [2, 3]. Whenever the DJJ is malpositioned and the cecum is mobile or located in the upper abdomen, there may be a narrow mesenteric base and accompanying risk of small bowel volvulus, justifying inclusion of these entities as malrotation [3].

Variants of normal or abnormalities of intestinal rotation are commonly associated with heterotaxy, both right and left isomerism. These include mirror image rotation, non-rotation and partial rotation/malrotation. Mirror image rotation, mirror image malrotation or mirror image non-rotation are simply the inverse of normal, malrotation or non-rotation, just associated with a right-side stomach and expected right-side DJJ [3].

Our purpose is to report our experience with heterotaxy and bowel rotation abnormalities by critically reviewing and correlating imaging studies, including newer modalities such as CT and MR imaging, with surgical findings to provide an approach to the management of these cases. For the purposes of simplification, rotation and mirror image rotation, malrotation and mirror image malrotation, and non-rotation and mirror image non-rotation are not differentiated in this study as they have the same appearance and significance, just inverted anatomy.

Materials and methods

We identified patients by undertaking a retrospective review of the heart center database from 1991 to 2013, searching for the terms heterotaxy, situs inversus, and right or left atrial isomerism to identify all cases. All cases so identified were confirmed to have heterotaxy by atrial or abdominal morphology. Approval for a retrospective chart review was granted by the Stanford Institutional Review Board.

Demographic and clinical information were obtained from the patient medical records. Imaging reports were reviewed from those children who had upper gastrointestinal (UGI) and/or small bowel follow-through (SBFT) fluoroscopic studies.

Imaging studies including initial and any additional UGI, SBFT, and CT or MR examinations that included the upper abdomen or abdomen/pelvis were then directly re-reviewed on the PACS system by a single pediatric radiologist (B.N.) who had access to the entire gamut of imaging studies, as well as the original reports, but was blinded to the surgical findings. On this re-review, when the DJJ was considered abnormal, a specific attempt was made to define the relative positions of the DJJ and cecum to infer the width of the mesentery. Cases with an abnormal DJJ and malpositioned or indeterminate cecum were classified as malrotation. Designation as non-rotation was reserved for those cases that had an abnormal DJJ, all of the small bowel on one side, most or all of the large bowel on the other, and cecum identified in the lower abdomen or pelvis suggesting a broad mesentery.

We obtained surgical reports from the medical chart and separately correlated them with both the original reports (UGI and SBFT studies) and the re-reviewed imaging findings (UGI, SBFT, CT and MR).

Results

Thirty-four heterotaxy patients were identified; 19/34 (56%) had an UGI at age 1 day-5 months. These 19 cases constitute the study group of children; 13/19 also had a concomitant SBFT. There were 15 males and 4 females, 9 had right isomerism (asplenia) and 10 had left isomerism (polysplenia). Six of the 19 children (32%) had specific gastrointestinal symptoms (vomiting in four, abdominal distention in two). Three children had nonspecific symptoms that could not definitely be attributed to the GI tract, such as difficulty in feeding or failure to thrive and the other ten were asymptomatic with GI studies obtained to screen for bowel rotation abnormality. All UGI/SBFT and CT/MR studies were obtained prior to any abdominal surgery with the exception of one child who presented with acute necrotizing enterocolitis and had bowel resection and a modified Ladd procedure prior to any imaging. CT scans that included the upper (2) or entire abdomen/pelvis (8) were obtained in 10/19 (53%) children; none of the CTs was requested to evaluate gastrointestinal abnormalities. Nine CT studies were obtained relatively early, at ages 1 day to 8 months, with one CT obtained at 3.5 years of age. While the position of the stomach and appearance of the liver and spleen were routinely mentioned in the formal radiology report, no mention was made of features relevant to bowel rotation in 7/10 cases, including superior mesenteric

artery/superior mesenteric vein (SMA/SMV) relationship, presence of retroperitoneal duodenum, position of small and large bowel and cecum. Two infants had an abdominal MRI, at 3 and 11 days of age respectively; the bowel was not mentioned in these reports either.

Practicing pediatric radiologists performed and interpreted all the initial fluoroscopic studies; the exact criteria used to differentiate malrotation from non-rotation were not specified. The fluoroscopy reports (including SBFT in 13/19) concluded that 4/19 (21%) were normal (Fig. 1), 15/19 (79%) abnormal, 4/19 (21%) were called non-rotation (including one case post modified Ladd procedure), and 11/19 (58%) malrotation (Figs. 2 and 3 and Table 1).

Subsequent re-review, with inclusion of two repeat UGI/SBFT studies as well as CT/MR findings, led to some changes with 2/19 (10%) designated as normal and 17/19 (90%) as abnormal, 9 with left and 8 with right isomerism, 7/19 (37%) classified as non-rotation, and 10/19 (53%) classified as malrotation (Figs. 1, 2, 3 and 4 and Table 2). In spite of obtaining SBFT studies, the position of the colon and cecum was indeterminate in 4/13 children on fluoroscopic examinations, most often due to termination of the study before the cecum was definitely identified (Fig. 2). There were no cases of volvulus although two children had emergent fluoroscopic studies (one was a repeat UGI) because of bilious vomiting.

Aside from clearly identifying the variant anatomy of the liver, hepatic vessels, spleen, stomach and pancreas (Figs. 1, 2, 3 and 4), the specific contributions of CT and MR in 12 children included: helping to identify normal versus abnormal bowel rotation by identifying the presence (1) or absence (10) of a retroperitoneal duodenum (Figs. 1, 2, 3 and 4); defining or confirming the location of small and large bowel (5) (Figs. 2, 3 and 4); defining the location of the cecum/appendix (3) (Figs. 3 and 4), and identifying normal (5) or abnormal (7) SMA/SMV relationships (Figs. 1, 2, 3 and 4, Table 3). Differentiating non-rotation from malrotation was only successful on CT/MR when the small bowel/large bowel and cecal positions were clearly identified (3 cases) (Figs. 3 and 4). Absence of retroperitoneal duodenum was seen in both non-rotation and malrotation (Figs. 2, 3 and 4), and a crossing retroperitoneal duodenum was clearly present in one child who originally was thought to have a normal UGI/SBFT but who had a clearly abnormal repeat study (Fig. 1); likewise, abnormal SMA/SMV relationship (SMV reverse of expected or directly anterior) was only present in conjunction with abnormal rotation but occurred in both non-rotation and malrotation (Figs. 2, 3 and 4). Normal SMA/SMV (SMV to the right and anterior, with left-side stomach, and to the left and anterior, with right-side stomach) was present in one child with normal bowel rotation as well as in four children with other features consistent with abnormal bowel rotation (Table 3).

Surgical decisions were based on the original radiology fluoroscopy reports with the four cases initially reported as normal and four as non-rotation having no surgery (Table 1). Nine of 11 patients, originally classified as malrotation, underwent elective surgical exploration (Table 1). One child died of other causes prior to elective surgery, and one was lost to follow-up. Four of the nine operative cases were found to have non-rotation with broad mesenteric fixation and no bands, and they had no further Ladd procedure. Five had classic malrotation (3) or partial malrotation (2) with a narrow mesentery and bands and underwent a Ladd or modified Ladd procedure (Table 1). There was, therefore, a discrepancy between UGI/SBFT and surgical findings in 4/9 (44%) cases; all had been designated as malrotation on UGI and/or SBFT but were found to have non-surgical non-rotation at surgery (Table 1).

Surgical correlation with the re-reviewed imaging findings only had a discrepancy in 1/9 (11%) cases, which was called malrotation because the DJJ was abnormal but the position of the cecum was uncertain; this child was found to have non-rotation with a long mesentery, low cecum and no bands at surgery (Fig. 2, Table 2). Three cases reclassified as non-rotation with a broad mesentery were confirmed surgically (Table 2). One child, post heart transplant, was initially called normal but reclassified as malrotation after a second UGI/SBFT because of feeding difficulty and is awaiting elective surgery (Fig. 1).

The surgical approach was laparoscopic in 8/9 cases; however, in one of these eight children, this was converted to an open procedure to accomplish a Nissen fundoplication that could not be performed laparoscopically because of the unusual position of the liver. One child had an open surgical procedure because of a presentation with necrotizing enterocolitis (NEC) unrelated to the malrotation. In 8/9 cases, additional surgical procedure/s were performed at the same time as the malrotation surgery; these included appendectomy (5), Meckel diverticulectomy (1), gastrostomy (2), fundoplication (1), umbilical hernia repair (1), resection of bowel for NEC (1) and tracheostomy (1). No surgical or perioperative complications occurred in the nine operative cases. Six of nine cases had no postoperative complications reported, and no postoperative symptoms were considered related to the malrotation surgery. Among the three children with postoperative problems, the child with NEC and bowel resection required postoperative total parenteral nutrition support and eventually died due to the severity of the underlying cardiac condition; an additional child's death, several months after the gastrointestinal surgery (with good recovery), was attributed to renal failure and underlying complex heart disease, and one child had ongoing feeding difficulty and symptoms attributed to gastroesophageal reflux that had also been present preoperatively. Other than the one child mentioned above (Fig. 1), none of the nonsurgical cases has presented with acute gastrointestinal symptoms.

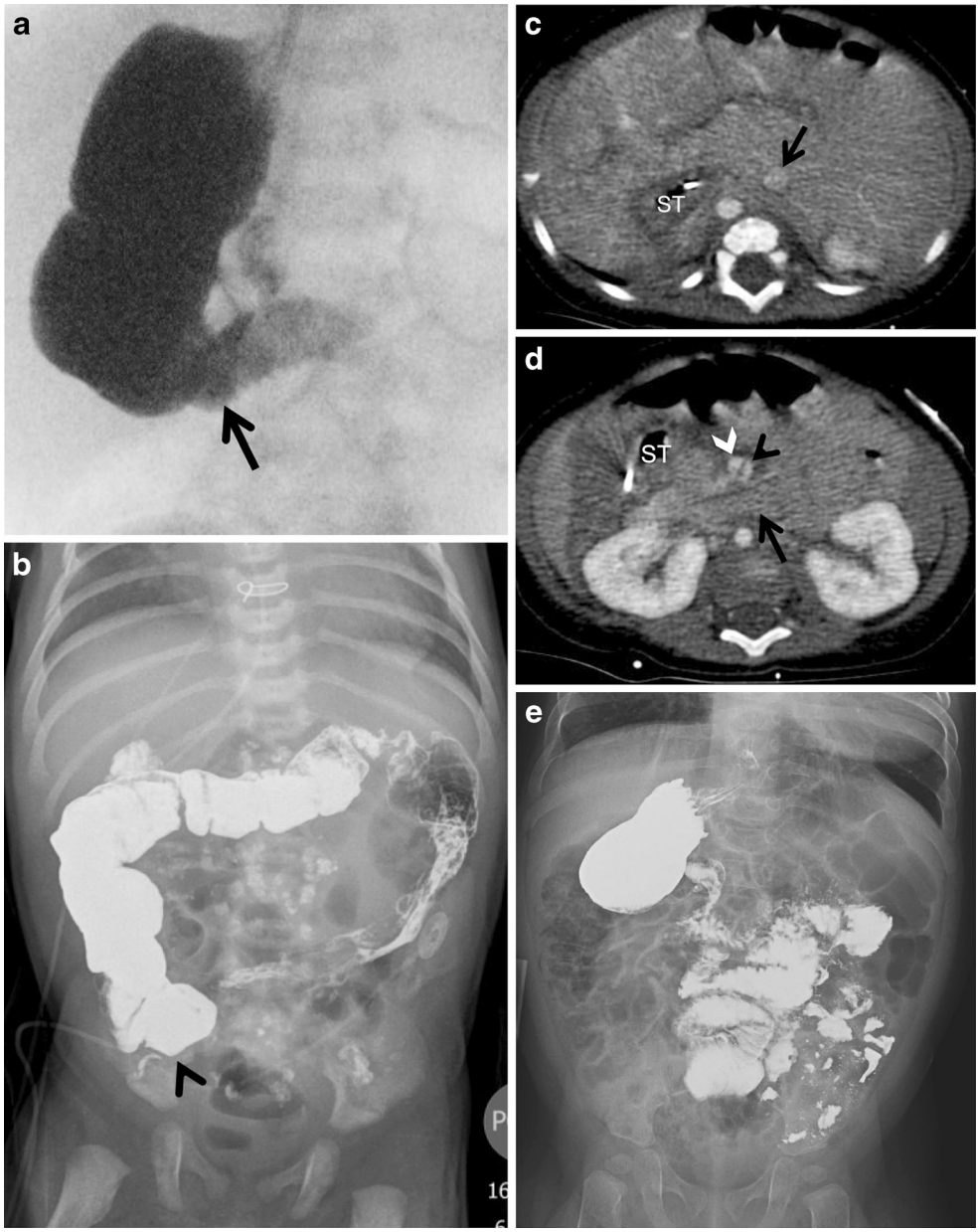

Fig. 1 Initial normal with agreement between initial fluoroscopy report and later combined UGI/SBFT plus CT reading but discrepant with later UGI/SBFT. Female with right isomerism. UGI/SBFT at 2 weeks of age (**a**). Right-side stomach with mirror-image position of the DJJ (*arrow*). **b** Delayed image shows normal colonic distribution with definitive location of the cecum and appendix in the right lower quadrant (*arrowhead*). One might argue that a right-side cecum is not normal in the presence of a right-side stomach. Nonetheless, the relative positions of the DJJ and cecum in this study suggest a broad mesenteric base. This also re-emphasizes that small and large bowel rotation are independent events. **c, d** Contrast-enhanced CT abdomen/pelvis axial scans, age 3 months, demonstrate a horizontal liver, absent spleen and right-side stomach (ST) with right-side abdominal aorta and left-side IVC (*arrow* in **c**). Note the presence of crossing retroperitoneal duodenum (*arrow* in **d**) passing between the aorta and SMA (*arrowhead* in **d**) with the SMV (*white arrowhead* in **d**) anterior and to the right of the SMA (abnormal with a right-side stomach). The position of the cecum could not be identified with certainty on this examination. **e** Repeat UGI/SBFT, age 9 months, post heart transplant, nonspecific abdominal symptoms. On this occasion, the DJJ is abnormally low with small bowel on both sides of the abdomen, indicative of malrotation. The cecum appeared higher than on prior study. The child is now scheduled for a laparoscopic Ladd procedure. This case emphasizes an important pitfall in the imaging assessment of malrotation. When the cecum is mobile, it may appear to be in normal position, with an erroneous assessment of a broad mesentery. In the presence of clinical symptoms or equivocal imaging findings, there should be a low threshold for repeat imaging

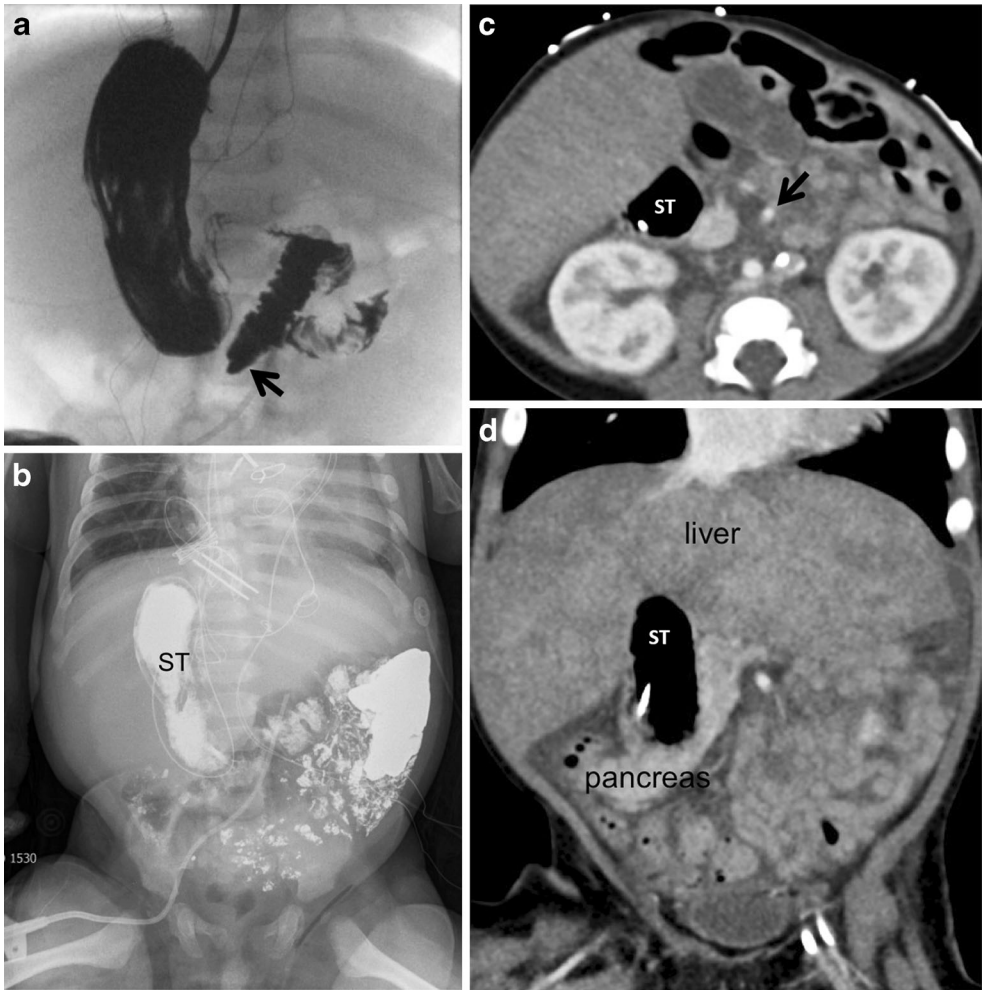

Fig. 2 Malrotation with agreement between initial fluoroscopy report and later combined UGI/SBFT plus CT reading, surgery discrepant with both, found non-rotation, broad mesentery, cecum right lower quadrant. Male with right isomerism. UGI/SBFT (**a, b**), at age 4 months, shows a right-side stomach with abnormal central and low position of DJJ (*arrow* in **a**). In spite of a 5-h delayed image, which showed all of the small bowel on the left (**b**), the cecum and colon location were not defined. Contrast-enhanced axial CT (**c**), age 3 months, and coronal reconstruction (**d**) demonstrate a right-side stomach (ST), horizontal liver, absent spleen, malpositioned pancreas, and left-side IVC. There is an absence of retroperitoneal duodenum between the SMA (*arrow* in **c**) and aorta. The SMV is anterior and to the right of the SMA (abnormal with a right-side stomach). On the coronal image (**d**), all of the small bowel appears left-side and all the colon right-side. However, this was designated malrotation because the position of the cecum was uncertain on both the fluoroscopic study and CT. This case reflects the complex issues associated with imaging in malrotation. In the light of imaging uncertainty regarding cecal position in the context of small bowel malrotation, laparoscopic surgical evaluation is not inappropriate

The 15/34 children with heterotaxy who did not have UGI studies had their clinical and imaging studies reviewed as a potential control group to compare with the study group. Four of the 15 were excluded from this group because of early neonatal death (3) from cardiovascular disease or early loss to follow-up (1) at age 3 months. Of the remaining 11 children, five (45%) had established malrotation (two with concomitant diaphragmatic hernias, two with Ladd procedures and one with surgery for necrotizing enterocolitis with concomitant Ladd procedure). The Ladd procedures were performed at outside facilities and the details of presentation, symptoms and anatomy are unknown. Three of the 11 had nonspecific GI symptoms treated by G-tube (3) and Nissen fundoplication (2), all at outside hospitals; 3/11 children (followed for 6-21 years) were reported to have no gastrointestinal symptoms. None of the children from this group of 15 had CT scans that included enough of the upper abdomen to assess bowel rotation; there were no abdominal MRIs. There was, therefore, incomplete data regarding the presence or absence of malrotation in this group of children.

Discussion

Many gastrointestinal abnormalities have been found in association with heterotaxy syndromes; these include biliary atresia, congenital diaphragmatic hernia, hiatal hernia, antral and duodenal web, annular pancreas, preduodenal portal vein,

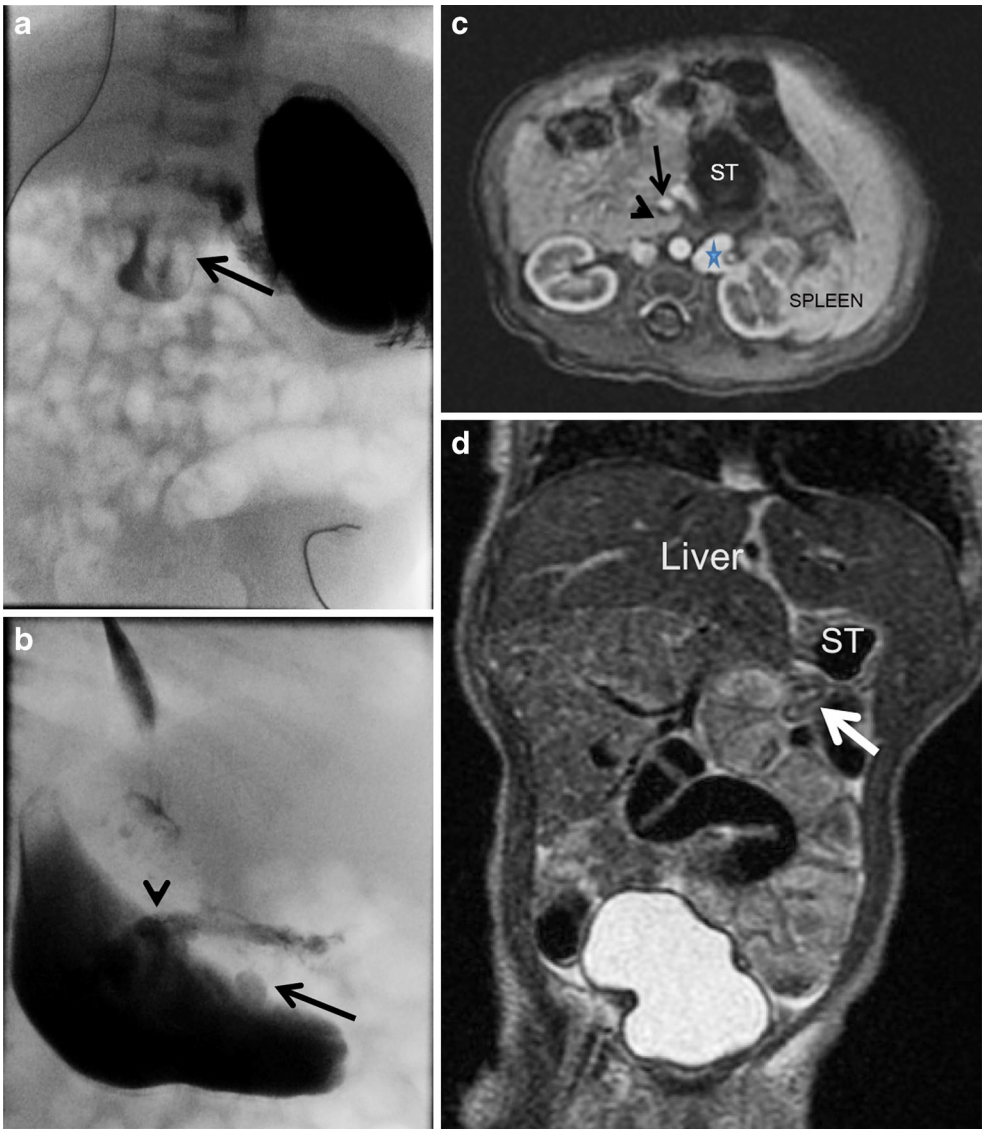

Fig. 3 Malrotation with agreement between initial UGI report and re-review plus MR. Correlates with surgery, partial rotation, short mesentery, bands, cecum left upper. Female with left isomerism. UGI (**a, b**) at age 10 days, no SBFT. Frontal (**a**) and lateral (**b**) fluoroscopic images demonstrate a left-side stomach with abnormal low and medial position of the DJJ, not crossing the spine (*arrow* in **a**) and located in front (*arrow* in **b**) of the duodenal bulb (*arrowhead* in **b**) and second portion of the duodenum indicative of abnormal bowel rotation. Axial MR post-contrast (**c**) and T2-W coronal MR (**d**) at age 10 days demonstrate horizontal liver, left-side stomach and left-side polysplenia as well as a left-side splenorenal shunt (*). There is an abnormal SMA/SMV relationship (SMV left and anterior to SMA (*arrow* in **c**). The duodenum (*arrowhead* in **c**) pokes in behind the SMA but does not cross the spine. On the coronal view (**d**), all of the small bowel appears to be right-side and the colon left-side, but the cecum and appendix are identified in the left upper quadrant (*arrow* in **d**), suggesting a narrow mesenteric base and risk for volvulus

splenorenal shunt, small bowel atresia, Meckel diverticulum, duplication cyst, Hirschprung disease and imperforate anus [7, 8]. Abnormal intestinal rotation is particularly common (40-90% in the literature, 90% in our study group and 45% in the control group [with incomplete data]) [3, 6]. Abnormal rotation was almost evenly distributed in left (9/10) versus right isomerism (8/9) in our study cases and malrotation was more commonly diagnosed by imaging (10/19, 53%) than non-rotation (7/19, 37%) with close surgical correlation (discrepancy in 1/9) (Table 2).

Children with heterotaxy frequently have many other medical problems, particularly complex congenital heart disease that greatly overshadows the question of malrotation. As more children survive cardiac surgery, other clinical problems, such as gastrointestinal abnormalities, need to be addressed. Only 6/19 in our study group had symptoms specifically attributable to the GI tract, but other clinical problems could mask GI symptoms or create confusion as to whether such symptoms as feeding intolerance or failure to thrive were related to a gastrointestinal or cardiorespiratory cause. Ten of the 19 in our study group had no reported gastrointestinal symptoms, eight of these patients had abnormal bowel rotation on imaging; 3/11 in the control group were asymptomatic. Although we had no cases of volvulus, they have been described [6, 9–11] with a potentially catastrophic outcome.

Our current practice recommendation is to include a US of the abdomen as part of the initial evaluation of a child with possible heterotaxy (Fig. 5). This can include evaluation of the liver, spleen, pancreas, vessels and bowel. We then image our patients electively with an UGI and SBFT as soon as they are reasonably medically stable (after stage 1 surgery in the case of a single ventricle repair) and inform the parents of the risk and symptoms of volvulus where appropriate (see algorithm, Fig. 5).

Table 1 Initial upper gastrointestinal series/small bowel follow-through (UGI/SBFT) fluoroscopic imaging reports correlated with surgical findings

Imaging findings: Initial		Surgical correlation
n=19	(UGI/SBFT)	
Normal	4 (21%)	No surgery - 4
Non-rotation	4 (21%)	No surgery - 4
Malrotation	11 (58%)	Elective surgery - 9
		• non-rotation, broad mesentery, no bands - 4
Volvulus	0	• partial malrotation with narrow mesentery, bands - 2
		• malrotation with narrow mesentery, bands - 3
		No surgery - 2
		• died of other causes before planned surgery - 1
		• lost to follow-up - 1
		UGI/SBFT/surgical discrepancy - 4
		• all non-rotation/broad mesentery at surgery

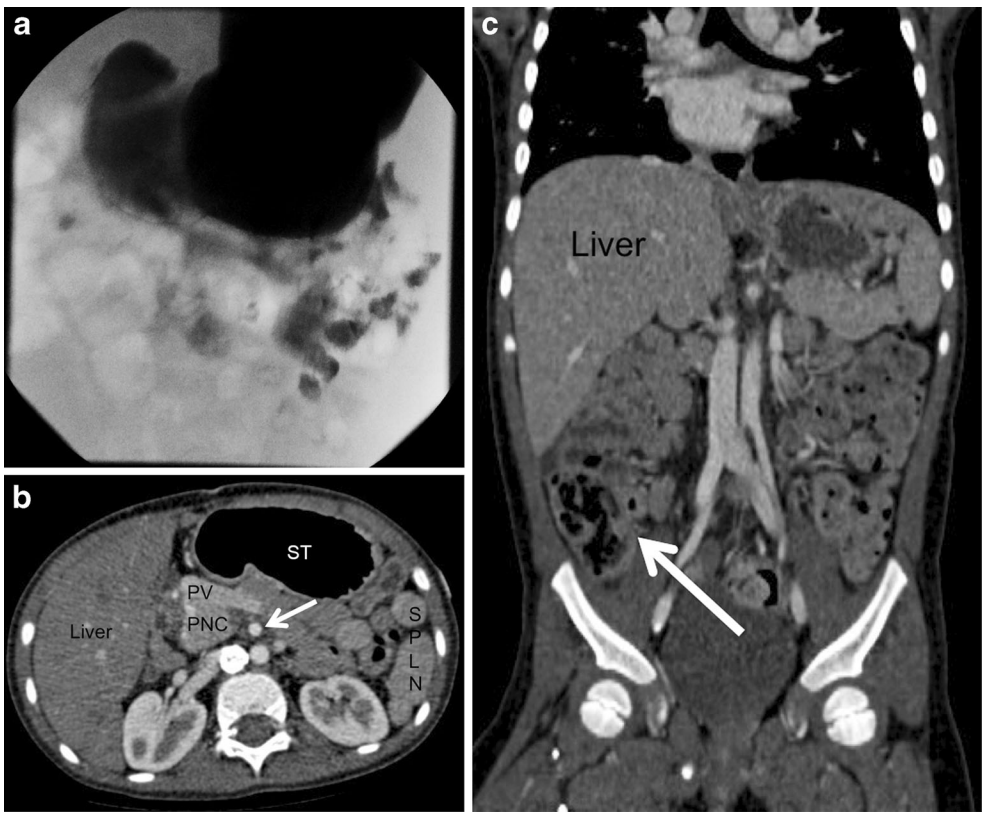

Fig. 4 Discrepancy between original fluoroscopy report (normal UGI) and CT (malrotation), no surgery. Female with left isomerism. UGI (**a**), age 3 months, the stomach was overfilled and the first pass of the contrast through the duodenum was not captured, making interpretation difficult. The original interpretation was considered normal. On re-review, the study was designated equivocal with a redundant duodenum and possible low DJJ. No surgery, based on original report. Contrast-enhanced CT, at age 3.5 years, axial (**b**) and coronal (**c**) images demonstrate a right-side liver, left-side stomach (ST) and polysplenia (SPLN). There is a preduodenal portal vein (PV) with the SMV extending anterior and to the left (abnormal) of the SMA (*arrow* in **b**). There is no retroperitoneal duodenum between the aorta and the SMA. The pancreas (PNC) is retroportal and probably annular. On the coronal view, the small and large bowel are located on both sides of the abdomen, and the cecum and appendix are in the right lower quadrant (*arrow* in **c**), suggesting that, although there appears to be small bowel malrotation, there may be a relatively broad-based mesentery. The child's older age at the time of the CT also suggests a lower likelihood of volvulus

Table 2 Re-review UGI/SBFT plus computed tomography/magnetic resonance (CT/MR) imaging findings correlated with surgical findings

Imaging findings: Later		Surgical correlation
n=19	(UGI/SBFT,CT,MR)	
Normal	2 (10%)	No surgery – 2
Non-rotation	7 (37%)	No surgery – 4
		Surgery – 3
		• Confirmed non-rotation with broad mesentery
Malrotation	10 (53%)	Elective surgery – 6
Volvulus	0	• Non-rotation, broad mesentery, no bands – 1
		• partial malrotation with narrow mesentery, bands – 2
		• Malrotation with narrow mesentery bands – 3
		No surgery – 4
		• Died of other causes before planned surgery – 1
		• Lost to follow-up – 1
		• 1 UGI/SBFT-CT discrepancy- no surgery
		• 1 abnormal on repeat UGI, awaiting surgery
		Imaging/surgical discrepancy – 1
		• non-rotation with broad mesentery at surgery

Table 3 CT/MR findings relevant to bowel rotation *n*=12

Retroperitoneal duodenum	Present-1 (malrotation)	Absent-10 (non/malrotation)
Defined location of small/large bowel	Yes-5	
Defined location of cecum/appendix	Yes-3	
SMA/SMV relationship	Normal-5 (4 w/malrotation)	Abnormal-7 (non/malrotaion)

SMA superior mesenteric artery
SMV superior mesenteric vein

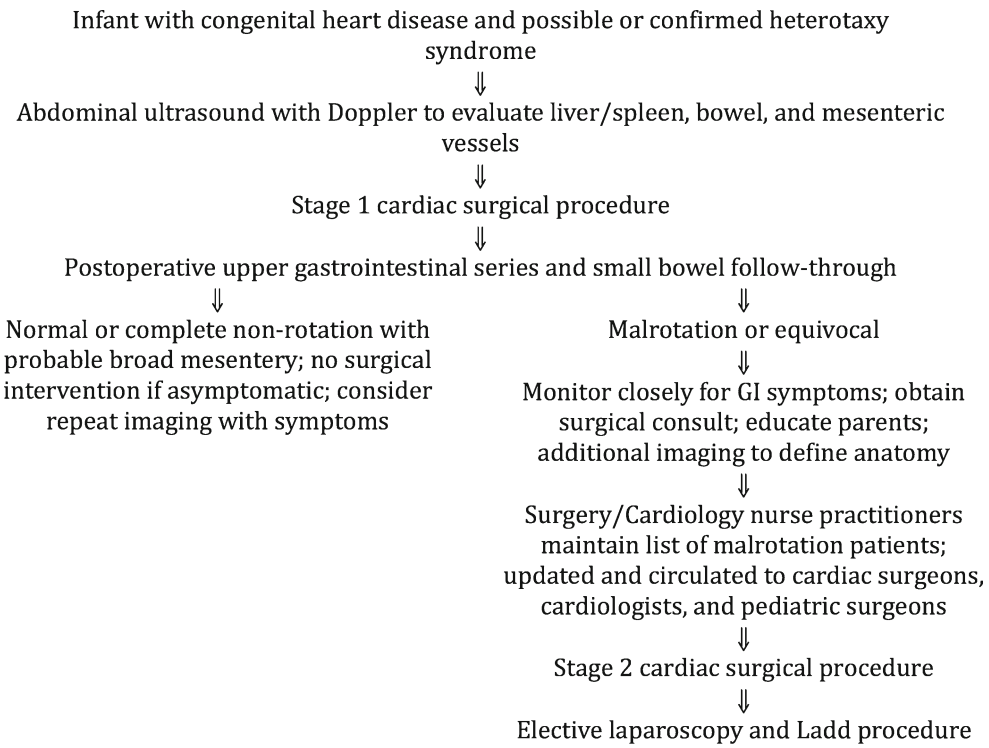

Fig. 5 Malrotation algorithm for patients with heterotaxy and complex congential heart disease

Elective surgical intervention in asymptomatic children with heterotaxy and malrotation is still somewhat controversial. Some reports have advocated a conservative approach in asymptomatic children because of the low incidence of volvulus and severe underlying disease (mostly cardiac) in many of these children with attendant high anesthetic/surgical risk, and reported higher incidence of postsurgical complications, including cardiorespiratory problems, sepsis and bowel obstruction [2, 8, 11]. Approximately two-thirds of cases of small bowel volvulus in both heterotaxy and non-heterotaxy malrotation cases occur in the first month of life and 90% in the first year [3, 11, 12], with the argument that, as long as there is close follow-up and prompt investigation of GI symptoms, asymptomatic children can be monitored without elective surgery [2]. Other reports indicate that the surgical and perioperative risks are not substantially higher than in non-heterotaxy cases and support elective surgical intervention rather than waiting for symptoms to appear, with a much higher presentation with volvulus in symptomatic children (potentially catastrophic) and more complicated postsurgical recovery [6, 9]. Surgery can now often be performed laparoscopically with low surgical morbidity and mortality, as reflected in our cases. The surgeons at our institution advocate elective laparoscopic surgery in heterotaxy cases with malrotation demonstrated on imaging studies even if patients are asymptomatic. The surgical timing in our cases was usually after stabilizing cardiac surgery (stage 2, bidirectional Glenn in the case of a single ventricle staged repair) (Fig. 5). Our cases had no intraoperative or perioperative complications. The observed postoperative problems (mostly remote) were attributed to the underlying disease rather than the surgical procedure. As evidenced by our cases, many of these children required multiple surgical procedures, such as gastrostomy, fundoplication and hernia repair, that could be performed concomitantly with a Ladd procedure.

The Ladd procedure consists of lysis of intra-abdominal adhesive bands in order to effectively broaden the mesenteric pedicle and widely separate the position of the small intestine and colon. The intended effect of the Ladd procedure is to recapitulate the position of the bowel encountered in non-rotation associated with a broad mesentery. When there is partial malrotation with a narrow mesentery, there are frequently adhesive bands that require lysis, and the cecum is repositioned in the lower abdomen to broaden the mesentery. This study confirms that there is a very high incidence of abnormal bowel rotation in heterotaxy, with 17/19 (90%) of our cases suggested on imaging and confirmed surgically in nine children. Separating non-rotation from malrotation is more complex. Our data suggest that this needs to be very carefully reviewed on imaging. It is not sufficient to demonstrate an abnormal DJJ because that is common to both malrotation and non-rotation. It is also insufficient to diagnose non-rotation based only on the position of the small and large bowel in the abdomen. It is important to convincingly demonstrate the location of the cecum when the DJJ is abnormal. A high cecum along with an abnormal DJJ with a short distance between the DJJ and cecum suggests malrotation with a narrow mesentery and attendant risk of volvulus. All cases with an abnormal DJJ and high, or indeterminate, cecum should be considered malrotation. Non-rotation should be reserved for those cases where the bowel distribution suggests non-rotation *and* the cecum appears low in location, as was determined in 7/19 (37%) of our re-reviewed imaging cases, suggesting that there may be a broad mesentery. Our study suggests that this designation of cases and careful evaluation of the cecum, using multiple imaging studies, if needed, lead to a better classification of patients, improved surgical correlation and the potential to avoid unnecessary surgery.

The approach of looking directly at the issue of a broad versus narrow mesentery helps avoid the confusion as to what exactly constitutes problematic bowel rotation and fixation in these very anatomically complex cases where the stomach, small bowel and large bowel are in variable positions. We advocate obtaining both an UGI *and* SBFT that continue long enough to identify the position of the cecum and colon in children with heterotaxy. When the DJJ is malpositioned, location of the cecum in the upper abdomen correlates with the highest risk for volvulus [13]. A low cecum could be a spurious finding at a single point in time, if the cecum is mobile; however, confirmation of a low cecum on more than one study is somewhat reassuring. Therefore, in a symptomatic child, there should be no hesitation in repeating the fluoroscopic study. A low cecal position lessens but does not eliminate the risk of volvulus as a small number of cases have been described in association with a fixed low position of the cecum [13]. Other imaging strategies that might be helpful include obtaining a contrast enema as well as correlation with CT and MR studies. Our cases indicate that careful attention to attempting to assess the width of the mesentery on imaging should result in somewhat improved diagnostic precision and help in avoidance of unnecessary surgery. Nonetheless, there may still be equivocal cases where laparoscopic exploration may be warranted (Fig. 2).

Reports in the literature suggest that the fluoroscopic assessment of malrotation can be difficult with as high as 15% false-positive and 6% false-negative rate caused by conditions such as dilated bowel, mass or organomegaly, and mobile DJJ and cecum [12]. It is essential to obtain both lateral and supine frontal views of the DJJ at fluoroscopy to confirm the posterior position of the fourth portion of the duodenum relative to the proximal duodenum as well as the normal position of the DJJ crossing the spine and ascending to the level of the gastric antrum [1, 3] (Fig. 3). Other authors have also suggested that the addition of a SBFT or enema, to confirm questionable abnormality, is helpful in difficult cases [1, 12, 13]. In some cases, in spite of an adequate SBFT and even a barium

enema, there may still be some uncertainty about the bowel position.

We are not suggesting that cross-sectional imaging be routinely obtained to assess for malrotation. However, US, CT and MR studies are often obtained for other reasons and can provide useful confirmatory or supplemental information regarding the overall visceral arrangement, as well as features of bowel rotation abnormalities, especially in cases that are confusing or equivocal on UGI/SBFT. The features of bowel rotation should be included in the evaluation and interpretation, of particular value is locating the small and large bowel and especially the location of the cecum/appendix. While the absence of a retroperitoneal duodenum and abnormal SMA/SMV relationship were readily identified and suggested an abnormality, they were not helpful in separating non-rotation from malrotation. Furthermore, similar to the experience reported by other authors, the SMA/SMV was normal in four of our patients with abnormal bowel rotation [4, 5]. Other studies have confirmed that identifying a retroperitoneal duodenum on CT is helpful in suggesting normal small bowel rotation. There is, however, a description of a case similar to our own where a retroperitoneal crossing duodenum seemed to be present in spite of clear malrotation on the UGI [4] (Fig. 1). Another author has pointed out that it is embryologically possible for a portion of the duodenum to be retroperitoneal when only 180° of small bowel rotation has occurred, and these cases are at risk for volvulus [14]. Ultrasound was not evaluated in the current study because it was impractical to do so in a retrospective imaging review. Ultrasound has been reported to accurately identify abnormal rotation [5]; the relevant anatomical relationships have to be specifically sought. Ultrasound would likely have similar shortcomings to CT/MR in separating malrotation from non-rotation.

Limitations of this study include its retrospective nature and relatively small numbers of patients with surgical correlation in 9/19 children.

Conclusion

Abnormal bowel rotation is common in patients with heterotaxy. Physicians taking care of these patients require a thorough knowledge of the spectrum of abnormality that can occur. We routinely obtain an early UGI and SBFT after initial surgical stabilization, with later elective laparoscopic surgery if needed. When the DJJ appears to be abnormal, the imaging approach should include visualizing the relative positions of both the DJJ and cecum to try to assess the width of the mesentery, a relative correlate for risk of volvulus, especially if there is a high cecum. Imaging by other modalities, including CT and MR, is often performed on these children, usually for unrelated reasons, but can provide useful supplemental information that increases diagnostic accuracy and confidence, as well as better stratification of appropriate surgical cases.

Conflicts of interest None

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