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Mimics of malrotation on pediatric upper gastrointestinal series: a pictorial review

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Abstract

Intestinal malrotation is a continuum of congenital anomalies due to lack of rotation or incomplete rotation of the fetal intestine around the superior mesenteric artery axis. The abnormal bowel fixation (by mesenteric bands) or absence of fixation of portions of the bowel increases the risk of bowel obstruction, acute or chronic volvulus, and bowel necrosis. The clinical presentation of patients with malrotation without, with intermittent, or with chronic volvulus can be problematic, with an important minority presenting late or having atypical or chronic symptoms, such as intermittent vomiting, abdominal pain, duodenal obstruction, or failure to thrive. The diagnosis is heavily reliant on imaging. Upper GI series remain the gold standard with the normal position of the duodenojejunal junction lateral to the left-sided pedicles of the vertebral body, at the level of the duodenal bulb on frontal views and posterior (retroperitoneal) on lateral views. However, a variety of conditions might influence the position of the duodenojejunal junction, potentially leading to a misdiagnosis of malrotation. Such conditions include improper technique, gastric over distension, splenomegaly, renal or retroperitoneal tumors, liver transplant, small bowel obstruction, the presence of properly or malpositioned enteric tubes, and scoliosis. All of these may cause the duodenojejunal junction to be displaced. We present a series of cases highlighting conditions that mimic malrotation without volvulus to increase the practicing radiologist awareness and help minimize interpretation errors.

Key words: Malrotation—Upper GI study—Duodenojejunal junction—Mimickers—Ligament of treitz

Intestinal malrotation is a continuum of congenital anomalies due to lack of or incomplete rotation of the fetal intestine around the superior mesenteric artery (SMA) axis [1–3]. The abnormal bowel fixation (by mesenteric bands) or absence of fixation of portions of the bowel increases the risk of bowel obstruction, acute or chronic volvulus, and bowel necrosis [4, 5]. Malrotation occurs in approximately 1 in 200–500 live births and is symptomatic in about 1 in 6000 live births [1, 2]. Most patients who are affected by malrotation show signs of the condition soon after birth with 75% of symptomatic cases diagnosed in the newborn period, and up to 90% of symptomatic cases diagnosed within the first year of life [3, 6, 7]. In newborns, malrotation with volvulus most commonly presents as bilious vomiting but it can also manifest as peritonitis, dehydration, shock or as bloody vomitus due to intestinal necrosis [2, 3, 8]. However, the clinical presentation of patients with malrotation without, with intermittent, or with chronic volvulus can be problematic, with an important minority presenting late or having atypical or chronic symptoms, such as intermittent vomiting, abdominal pain, duodenal obstruction, or failure to thrive [7]. In asymptomatic patients, the diagnosis of intestinal malrotation may be incidental and the need for surgery is questionable. In particular, "non-rotation," a variant defined by small bowel located in the right side of the abdomen and colon located in the left, has a much lower risk of volvulus [3].

In both the typical and atypical presentations, the diagnosis is heavily reliant on imaging [9]. The current gold standard for diagnosis of rotation abnormalities is upper gastrointestinal contrast radiography (UGI) [1, 10, 11]. The sensitivity of UGI for the diagnosis of malro-

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tation is reported between 93% and 100% [5, 6, 12, 13]. However, there is an approximately 15% rate of false-positive UGI examinations [5].

During UGI exams, the normal position of the duodenojejunal junction is lateral to the left-sided pedicles of the vertebral body, at the level of the duodenal bulb on frontal views and posterior (retroperitoneal) on lateral views [5, 14, 15]. However, in order to appropriately identify and use these anatomic landmarks, close attention to technique and positioning must be paid. Inappropriate positioning and rotation of the patients might easily lead to a false-positive result [16, 17]. Regarding positioning, for example, a leftward rotated patient (i.e., incompletely flat on the table) would artifactually separate the duodenojejunal junction from the underlying pedicles, simulating malrotation. Errors in positioning are more common when patients are excessively bundled, or in older children, when there is excessive motion. Obliquity can also make images harder to interpret correctly. Applegate et al. [5] described technical points to avoid false positive such as observing the first passage of contrast through the duodenum, avoiding overfilling the stomach and demonstrating the position of duodenojejunal junction both in frontal and lateral projections.

After proper adherence to imaging protocol, if the duodenojejunal junction is not to the left of the left-sided pedicles and retroperitoneal in course, a diagnosis of rotation abnormality should be made. However, inferior displacement of the normal duodenojejunal junction is a common variation seen on frontal views in infants, more common in premature infants, due relatively mobile ligament of Treitz [5]. In addition, false-positive exams can be due to a wide range of conditions, including gastric over distension, splenomegaly, renal or retroperitoneal tumors, liver transplant, small bowel obstruction, heavy enteric tubes pulling down the ligament of Treitz, malpositioned enteric tubes, and scoliosis. All of these may cause the duodenojejunal junction to be medially or inferiorly displaced [1]. We offer a series of cases highlighting conditions that mimic uncomplicated malrotation (i.e., without volvulus) to increase the practicing radiologist awareness and help minimize interpretation errors.

Discussion of cases

Case 1

Suggestion of malrotation due to an over distended stomach: A 2-year-old boy presented as an outpatient with history of frequent non-bilious emesis (Fig. 1). Initial UGI images showed marked gaseous distention of the stomach and abnormal orientation of the duodenojejunal junction and proximal jejunum, suggestive of malrotation. Marked gastric distention displacing the distal duodenum medially and inferiorly has been described as a mimicker for malrotation [3]. Additional images with a decompressed stomach showed normal position of duodenojejunal junction. At times, it might be necessary to insert a nasogastric tube in order to decompress the stomach before proceeding with contrast administration. In addition to helping better characterize the position of the duodenojejunal junction, a decompressed stomach helps with using less contrast and avoiding emesis during the exam.

Case 2

Mass effect from large left multicystic dysplastic kidney: A 4-month-old boy with multiple episodes of abdominal distention and feeding intolerance (Fig. 2). UGI images showed duodenum and duodenojejunal junction located on the right upper abdomen and to the right of the spine. However, the patient had a history of large left multicystic dysplastic kidney, which can be seen in the scout radiograph. Positive left retroperitoneal force pushing the duodenojejunal junction to the contralateral side has been described as a mimicking of malrotation [19].

Case 3

Mass effect from splenomegaly: An 8-year-old girl with portal hypertension presented with non-bilious vomiting (Fig. 3). The duodenojejunal junction was more medial in position than normal on UGI series. However, abdominal radiograph and MRI showed enlarged spleen displacing the bowel medially. Similar to retroperitoneal masses, left intraperitoneal mass effect (from transplanted left lobe of the liver or splenomegaly) can displace the duodenojejunal junction rightward [18, 19]. Partial hepatectomy, right nephrectomy, or congenital absence of the right kidney results in the creation of a potential space in the right side of the abdomen that allows bowel to migrate upward and rightward, also resulting in rightward displacement of the duodenojejunal junction. Misinterpretation can usually be avoided by a careful review of the scout image and/or previous imaging. In the absence of prior imaging, knowledge of prior clinical and surgical history is key.

Case 4

Mass effect from midline transplanted liver and potential space from the right hepatectomy defect: A 5-year-old male with neurologic impairment and history of mitochondrial disease status post liver transplant (Fig. 4). UGI study was requested due to concern for intermittent bowel obstruction. UGI images showed no evidence of bowel obstruction. There was, however, rightward and upward migration of the proximal duodenum, making the duodenojejunal junction low by comparison with also abnormal position of the majority of the small bowel and colon in the right hemiabdomen mimicking malro-

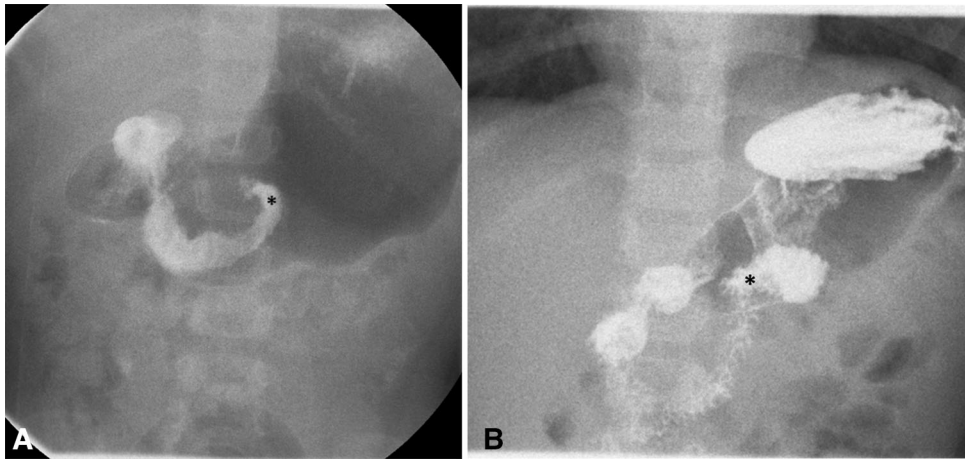

Fig. 1. Case 1: Gastric distension. A 2-year-old boy presented with history of frequent non-bilious emesis. Initial UGI images showed marked gaseous distention of the stomach with low positioning of the duodenojejunal junction (*) without clearing the left pedicle of L2 and exaggerated by mild rotation of the patient toward the left, suggestive of malrotation (**A**). Additional images with a decompressed stomach and in a straight supine position showed normal position of duodenojejunal junction (*) (**B**).

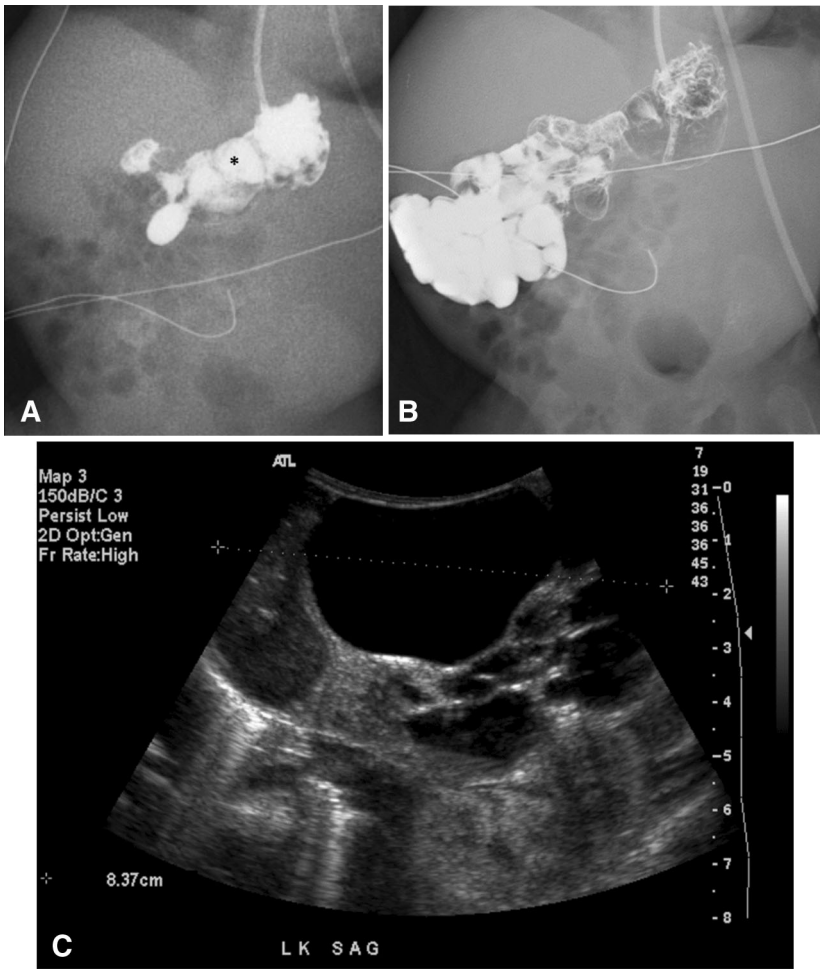

Fig. 2. Case 2: Mass effect from large left retroperitoneal masses. A 4-month-old boy with multiple episodes of abdominal distention and feeding intolerance. UGI images showed duodenum and duodenojejunal junction located to the right of the spine (*) (**A**). Noticed was mild rotation of the patient, which also was mildly affecting the apparent location of the duodenojejunal junction. Delayed images also showed the entire small bowel shifted to the right hemiabdomen due to mass effect from the large left retroperitoneal mass (**B**). Prior grayscale ultrasound image demonstrating the large left multicystic dysplastic kidney, measuring over 8 cm in length (**C**).

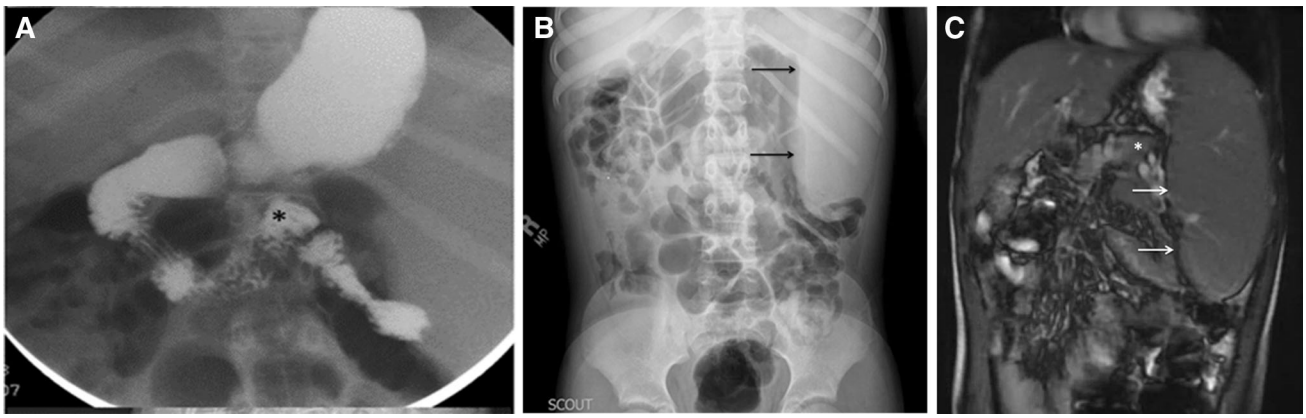

Fig. 3. Case 3: Mass effect from splenomegaly. An 8-year-old girl portal hypertension presenting with non-bilious vomiting. The duodenojejunal junction was more medial in position than normal on UGI series (*) (**A**). However, abdominal radiograph (**B**) and coronal Steady-State MR image (**C**) showed the enlarged spleen (arrows) displacing the duodenojejunal junction (*) medially, similar to retroperitoneal masses, avoiding misinterpretation.

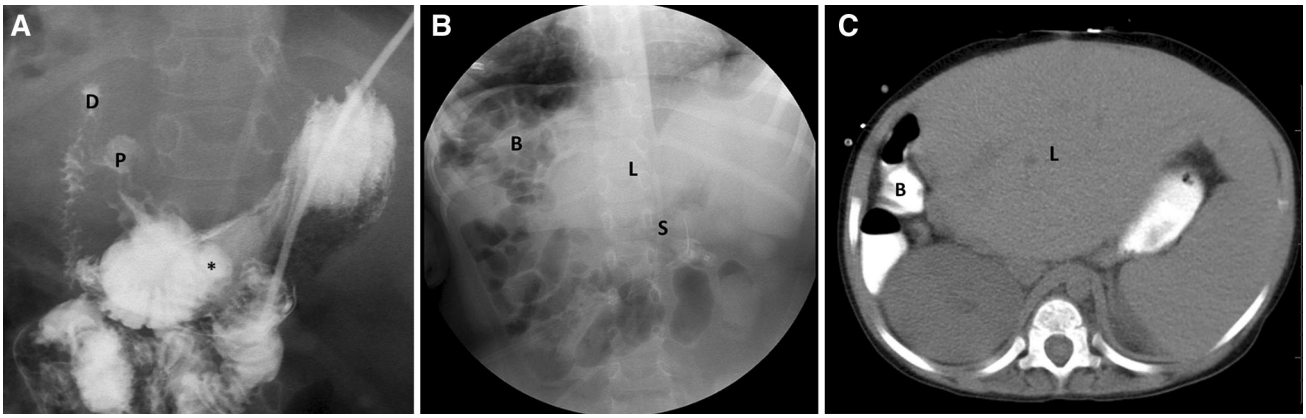

Fig. 4. Case 4: Mass effect from midline transplanted liver and negative force from the right hepatectomy defect. A 5-year-old boy with history of mitochondrial disease after liver transplant presenting with concern for intermittent bowel obstruction. UGI images showed rightward and upward migration of the pylorus (P) and the first and second portions of the duodenum (D), making the duodenojejunal junction (*) low by comparison and mimicking malrotation (**A**). Segmental liver transplantation is associated with distinctive anatomical distortion with the midline liver shadow (L), mass effect over the stomach (S), and right- and upward shifting of the bowel (B) to fill the hepatectomy defect (**B**). Axial CT image of the upper abdomen shows the comparatively larger midline transplanted liver (L) and the shifted small bowel (B) in the right upper quadrant (**C**).

tation. Segmental liver transplantation is associated with anatomic distortion of the course of the small bowel due to partial or complete hepatectomy and resultant disruption of normal bowel mesenteric and retroperitoneal fixation. There is often a characteristic imaging appearance, including migration of the duodenum and cecum into the hepatectomy fossa in the right upper quadrant [18].

Case 5

Pseudomalrotation due to chronic bowel dilatation: A 1-day-old infant admitted to the neonatal intensive care unit with prenatal diagnosis of jejunal atresia and now with bilious vomiting (Fig. 5). UGI series showed duodenojejunal junction at a lower level than expected, suspecting malrotation and confirming the diagnosis of jejunal atresia. However, the lateral view shows a normal retroperitoneal course of the duodenum. It is known that long-standing intestinal distention may cause inferior displacement of the duodenojejunal junction, thereby mimicking incomplete rotation of the midgut, and abnormal mesenteric fixation [20]. The patient underwent surgery, and intraoperative findings revealed type IIIB jejunal atresia (apple-peel appearance of the atretic bowel coiling around the ileocolic artery) with normally rotated bowel. This presentation can also be found in more distal bowel obstructions/atresias, in which patients might undergo UGI for initial evaluation of bilious vomiting. Hirschsprung

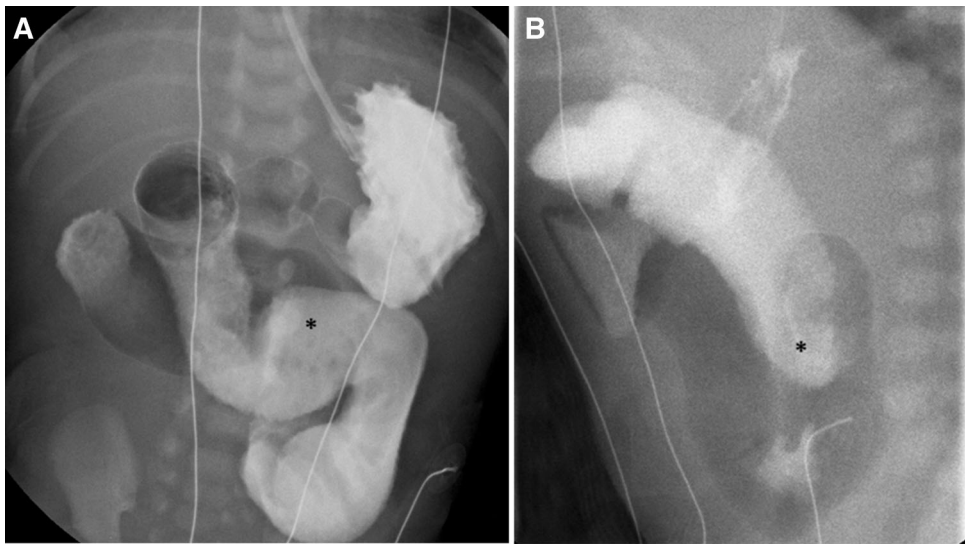

Fig. 5. Case 5: Pseudomalrotation due to chronic bowel dilatation. A 1-day-old boy admitted with prenatal diagnosis of jejunal atresia and now with bilious vomiting. UGI series showed the duodenojejunal junction (*) at a lower level than expected, suspecting malrotation and confirming the diagnosis of jejunal atresia (**A**). However, the lateral view shows a normal retroperitoneal course of the duodenum (**B**). The patient underwent surgery, and intraoperative findings revealed type IIIB jejunal atresia (apple-peel appearance of the atretic bowel coiling around the ileocolic artery) with normally rotated bowel.

disease, meconium ileus, and ileal atresia usually present with numerous dilated bowel loops [4, 21, 22]. These dilated loops may push down on a lax but otherwise normal ligament, leading to a misinterpretation of malrotation.

Case 6

Pseudomalrotation due to chronic distal bowel obstruction: A 2-week-old girl with history of ileal atresia and meconium pseudocyst status post ileostomy with mucous fistula (on day 1 of life), now with projectile bilious emesis (Fig. 6). An UGI study performed for suspected malrotation and midgut volvulus showed abnormally low position of the duodenojejunal junction, suggestive of malrotation without midgut volvulus. However, no malrotation was noted in the original surgical report and the patient improved without intervention. Subsequent UGI study 2 weeks later (now at 1 month of life) showed a return of the duodenojejunal junction to normal posi-

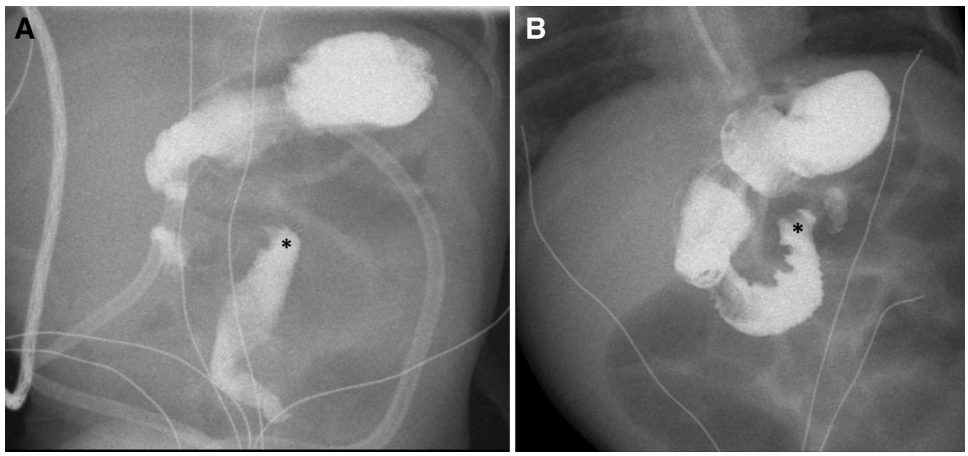

Fig. 6. Case 6: Pseudomalrotation due to chronic distal bowel obstruction. A 2-week-old girl with history of ileal atresia and meconium pseudocyst status post ileostomy with mucous fistula (on day 1 of life), presented with new projectile bilious emesis. An UGI study performed for suspected malrotation and midgut volvulus showed abnormally low position of the duodenojejunal junction (*) (**A**), suggestive of malrotation without midgut volvulus. However, no malrotation was noted in the original surgical report and the patient improved without intervention. Subsequent UGI study 2 weeks later (now at 1 month of life) (**B**), performed for confirmation of normal rotation, showed a return of the duodenojejunal junction (*) to normal position.

tion. This case further illustrates the challenges of interpreting UGI studies in patients with ongoing or (as in this case) recent chronic distal obstruction. Any obstruction past the ampulla could present with bilious emesis (even with imperforate anus or Hirschsprung disease, if there are enough bouts of vomiting) and the dilatation of proximal bowel loops weighs on and displaces the ligament of treitz downward (see comment about ligamentous laxity in case 7). While the dilatation of the bowel resolves promptly, the downward displacement of the ligament and its laxity takes longer to normalize. A careful review of the history with special emphasis of the surgical findings can avoid unnecessary subsequent procedures. However, after ruling out the presence of acute complications such as volvulus, a repeat examination up to 4 weeks after initial surgery might be needed for confirmation that the low lying duodenojejunal junction is due to prior chronic obstruction and not to true malrotation.

Case 7

Laxity of the ligament of Treitz: A 1-month-old term baby with esophageal atresia with tracheoesophageal fistula post repair (Fig. 7). Abdominal radiograph showed an unusual course of enteric tube, suggesting malrotation. After retracting the enteric tube, UGI series showed no malrotation. The course of the enteric tube suggesting malrotation was a consequence of the chronic indwelling line causing laxity of the ligament of Treitz. In infants with enteric tubes in the duodenum or jejunum, the weight of the tube may displace the normal duodenojejunal junction by stretching caudally the relatively lax ligament of Treitz [5]. Stretching of duodenal loop causing duodenojejunal junction misplacement can be corrected by retracting the enteric tube proximal to the third portion of the duodenum. Other long-standing tubes such as gastrojejunostomy tubes can have a similar effect over the ligament of Treitz and, similarly, retraction of the tube during UGI examination is a key to properly identify this mimicker.

Case 8

Malpositioned tube mimicking malrotation: A 2-year-old victim of child abuse and bowel injury who developed duodenal stenosis (Fig. 8). Initially malpositioned enteric tube, coiling in the third portion of a massively dilated duodenum, mimicked a low duodenojejunal junction. However, previous CT showed the dilated duodenum with normal rotation. Final image showed the tube properly positioned past the stricture into the normally located duodenojejunal junction.

Case 9

Malpositioned tube mimicking malrotation: An ex-28-week preterm with respiratory distress due to surfactant deficiency (Fig. 9). An enteric tube looped in the stomach and crossed the pylorus before terminating over the left S1 pedicle, suspecting malrotation. However, there was intra-abdominal free air depicted on AP view, confirmed in cross-table lateral view. The lateral radiograph can help to confirm the unusual retroperitoneal location and straight course of the tube and more readily identify the free air, which is most suggestive of perforation and not malrotation. In this situation, water soluble contrast can

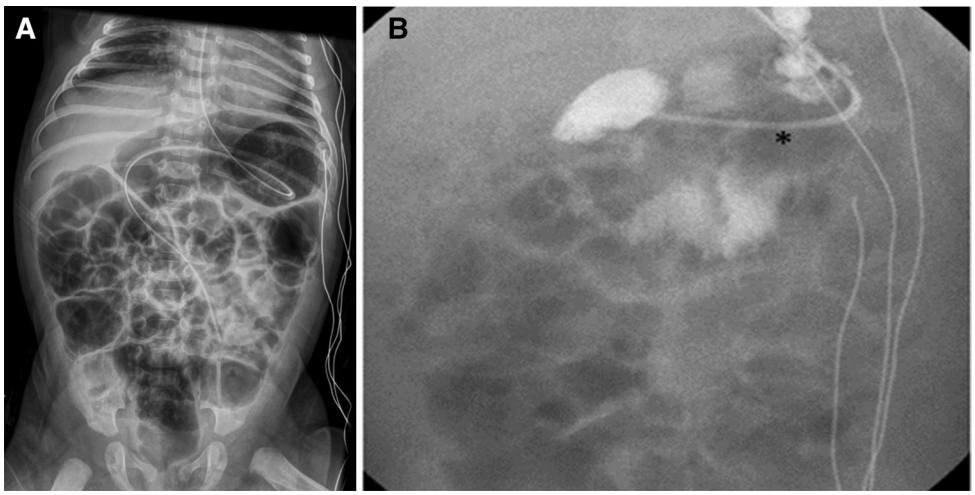

Fig. 7. Case 7: Laxity of the ligament of Treitz. A 1-month-old term girl with repaired esophageal atresia and tracheoesophageal fistula. Abdominal radiograph showed an unusual course of the enteric tube terminating in the left lower quadrant, suggesting malrotation (**A**). After retracting the enteric tube, UGI series showed no malrotation with normal duodenojejunal junction (*) (**B**). Stretching of duodenal loop causing duodenojejunal junction misplacement can be corrected by retracting the enteric tube proximal to the third portion of the duodenum.

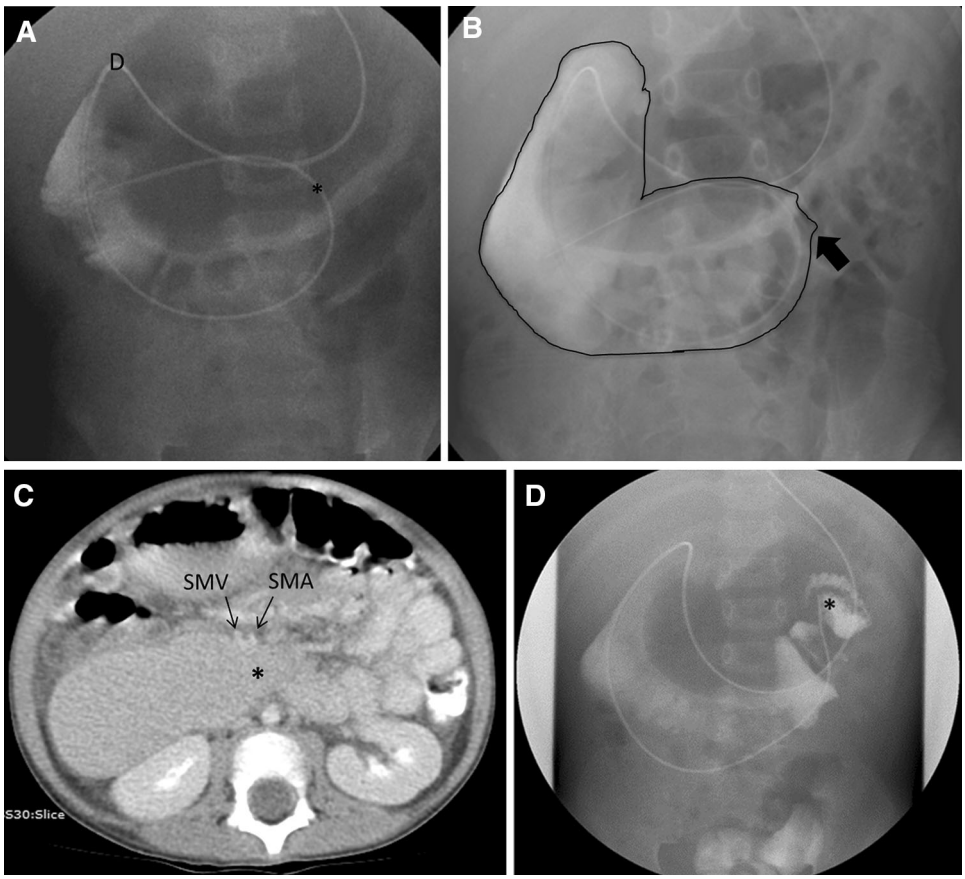

Fig. 8. Case 8: Malpositioned tube mimicking malrotation. A 2-year-old boy victim of child abuse and bowel injury who developed duodenal stenosis. Initially malpositioned enteric tube, coiling in the third portion of a massively dilated duodenum, mimicked a low duodenojejunal junction (*) compared to the first portion of the duodenum (D) (**A**). **B** outlines the duodenal contour. Previous axial contrast enhanced CT (**C**) showed the dilated duodenum (*) passing posterior to superior mesenteric artery and vein. Final image (**D**) showed the tube properly positioned past the stricture into the normally located duodenojejunal junction (*).

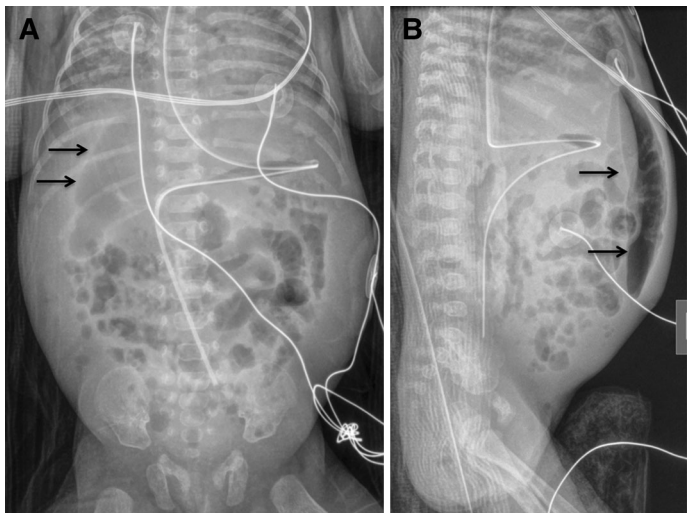

Fig. 9. Case 9: Malpositioned tube mimicking malrotation. An ex-28-week preterm girl with respiratory distress due to surfactant deficiency. An enteric tube looped in the stomach and crossed the pylorus before terminating over the left S1 pedicle, suspecting malrotation (**A**). However, there was intra-abdominal free air (arrows). It was confirmed in cross-table lateral view (**B**) (arrow). The patient underwent exploratory laparoscopy which revealed NG tube perforating the second portion of the duodenum with normal bowel rotation. The duodenum was repaired.

be used to further confirm the diagnosis if necessary. The patient underwent exploratory laparoscopy which revealed NG tube perforating the duodenum with normal bowel rotation. The duodenum was repaired.

Case 10

Scoliosis: An 11-year-old male with multiple medical problems and severe scoliosis, on gastrojejunostomy tube feeds (Fig. 10). From the UGI study, evaluating the

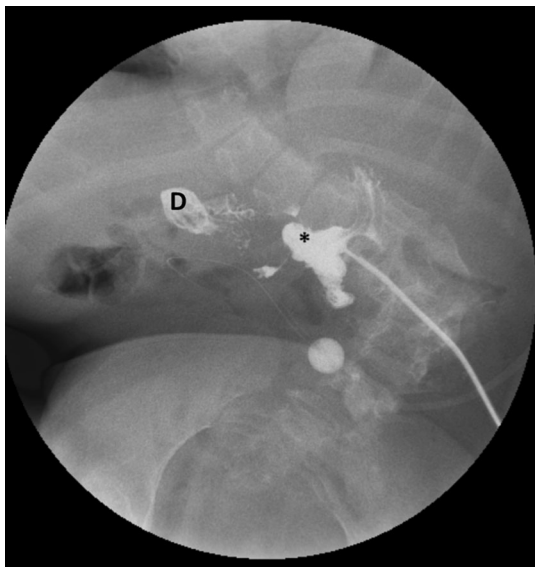

Fig. 10. Case 10: scoliosis. An 11-year-old male with complex medical history and severe scoliosis, with gastrostomy and jejunostomy tubes feeds. From the UGI study, the general configuration of the duodenal C-loop is normal with the duodenojejunal junction (*) to the right and at the level of the duodenal bulb (D). However, usual anatomic landmarks including the left pedicles cannot be used.

location of the ligament of Treitz is limited with the given severe scoliosis. Even though the general configuration of the duodenal C-loop is normal, the usual anatomic landmarks including the left pedicles cannot be used. Evaluating the position of ligament of Trietz in patients with scoliosis is challenging. Scoliosis may cause the duodenojejunal junction to be medially or inferiorly displaced and relationship to the spine is not reliable [1].

Discussion

Intestinal malrotation, when presenting with either intermittent or chronic symptoms, requires accurate imaging recognition for early surgical correction [5, 23]. The imaging diagnosis of intestinal malrotation is based on several factors, including the location of the duodenojejunal junction and the third portion of duodenum coursing retromesenteric, which can be made with ultrasound, computed tomography (CT), or magnetic resonance imaging (MRI) [24]. Special attention has been paid to ultrasound and MRI because of lack of radiation [10, 19, 25–28]. Ultrasound has shown very high sensitivity (93.8%–100%) and specificity (100%) with few indeterminate cases [28–31], while Fay et al. report confirmation of normal bowel rotation on 54%–64% of MRIs performed for non-related indications [10]. However, in spite of its use of ionizing radiation, UGI remains the gold standard [10, 11]. Current fluoroscopic technology can reduce the effective dose of most UGI examinations to less than 1 mSv [11].

When UGI studies are used to depict the position of the duodenojejunal junction and hence the presence or absence of malrotation, the interpreting radiologist must be aware of normal variations and other pathologies mimicking malposition of the duodenojejunal junction [1, 5, 19, 26]. The normal variants include (a) isolated finding of the jejunum in the right upper quadrant, (b) a normal-appearing duodenal sweep with the duodenojejunal junction over the left pedicle but not necessarily completely to the left of the spine, (c) proximal redundancy of the duodenum or duodenum mobile, and (d) clockwise rotation of the distal duodenum (duodenum inversum) [32]. A repeat UGI series with greater attention to documentation of the duodenal position or insertion of nasogastric tube to control the influx of barium can improve success but is rarely needed [26, 32]. Lim-Dunham et al. reported that manual epigastric compression during the UGI study allows an increased level of confidence in making the diagnosis of malrotation and volvulus [33]. Diagnostic confidence and accuracy may increase with a small bowel follow-through examination to depict the location of the small bowel and cecum. The cecum is abnormally positioned in 80% of patients with malrotation [5, 32]. While cecal position on contrast enema was originally performed to assess for malrotation, it is the location of the duodenojejunal junction that is considered the gold standard as normal cecal position does not exclude the diagnosis of malrotation [3].

We presented several cases of malrotation mimickers that can usually be suspected during the study acquisition or at the time of interpretation. For example, a gas-dilated stomach can be easily decompressed with a nasogastric sump/tube. A history of liver transplant, absent left kidney, or marked splenomegaly can also be obtained from the medical record or referring physicians before the test or be suspected on scout radiographs at the time of the study. Pulling of the ligament of Treitz by heavy enteric tubes or dilated bowel from distal obstruction can also be easily inferred from initial scouts, previous imaging and, of course, the presence of a transpyloric jejunal tube. Redundant duodenum and negative pulling force in right hemiabdomen causing displacement of duodenojejunal junction might be difficult to diagnosis if the malrotation is true or not. It is essential to obtain both lateral and supine frontal views of the duodenojejunal junction 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 duodenojejunal junction crossing the spine and ascending to the level of the gastric antrum [25]. At last, it is crucial to keep in mind that patients with malrotation mimickers (e.g., previous surgeries, indwelling catheters, or scoliosis) can also have a real malrotation. Clinical context should be considered as a priority. Surgery may still be warranted if symptoms

persist/worsen or are classic for malrotation with volvulus, such as bilious emesis or clinical signs of necrotic bowel.

Conclusion

We reviewed the mimickers of malrotation as seen on UGI series. To avoid unnecessary radiation and a high rate of false-positive studies, radiologists must be aware of underlying clinical scenarios that suggest malrotation in patients with normal rotation. Radiologists caring for children should be familiar with these potential pitfalls in which greater attention to technique, prior imaging and prior clinical history, can be helpful.

Compliance with ethical standards

Conflict of interest None.

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