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REVIEW

The position of the duodenojejunal junction: the wrong horse to bet on in diagnosing or excluding malrotation

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Abstract
Purpose The purpose of this communication is to highlight the shortcomings of all currently used imaging criteria in diagnosing or excluding malrotation and offer ultrasound demonstration of the 3rd portion of the duodenum (D3) between the AO and the SMA in transverse and sagittal plains as the most reliable diagnostic method.
Background Although UGI is currently considered to be the imaging modality of choice in diagnosis of malrotations, numerous publications indicate that in certain patients, false positives and negatives can be encountered.
Materials and Methods The material consists of more than 10 years experience in university settings, during which the author has used US as the definitive imaging modality for the work-up of malrotation. High resolution linear transducers (5-17 MHz) are the transducers of choice.
Imaging Plains:
A. Transverse

With gradual grading compression, the following landmarks are illustrated in cehaplocaudad directions in the fallowing order.

- The junction of splenic vein with the SMV portal vein.
- The cross sections of SMA and SMV that may either be situated in midline, or to the right or the left of the midline.
- Left renal vein crossing the spine from left to right between the AO and the SMA.
- The jejunal vein, often coming from left, transversing between the AO and the SMA.
- The transverse portion of the duodenum, D3, between the AO & the SMA

B. Sagittal

- D3 between vertically oriented SMA-SMV and the AO.
- If SMA is not aligned with the AO by slight compression on the right or the left side of the abdomen, it will be aligned (depending on leftward or rightward position of SMA-SMV in axial plain).
- Vertical orientation of SMA and SMV if they have an anteroposterior orientation.

C. Coronal

- Side by side orientation of SMA and SMV if they don't have an anteroposterior orientation.

Results In overwhelming majority of cases, by illustrating a retromesenteric D3 malrotation and, therefore, midgut volvulus were excluded.
Discussion None of the current imaging criteria addresses the following most fundamental anatomic and embryologic facts regarding the gut rotation and fixation.

- In first trimester, the D3 is secured in retroperitoneal space after the embryologic journey comes to an end, making the duodenum immune of midgut volvulus.
- The surgical pathology of malrotation-midgut volvulus indicates that D3 is always Intraperitoneal and has not reached its final embryologic destination in retroperitoneal space.
- Demonstrating a retromesenteric D3, therefore, indicates that the embryologic journey is completed and the patient does not have malrotation.
- Excluding malrotation excludes the likelihood of midgut volvus.

Conclusion The position of the DJJ, the configuration of the duodenal sweep, the orientation of the mesenteric

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vessels are all wrong horses to bet on because none of them addresses the fundamental anatomic and embryologic facts. Only the cross-sectional imaging, US, CT and MRI can prove that the D3 is retromesenteric, therefore, excluding malrotation and volvulus. Therefore, demonstrating a retromesenteric duodenum is the reference standard of imaging in the work-up of malrotation, not any other previously published criteria. The US imaging is the most acceptable imaging method for malrotation work-up, in the spirit of ALARA principle and "Image Gently" campaign.

Keywords CT · Malrotation · Midgut volvulus · MR and US

Background

Various imaging modalities have been used for diagnosis of malrotation-valvulus, including barium enema, UGI, ultrasound, and CT [1–28]. Some surgeons recommend laparoscopic diagnosis based on encountering enough cases in which imaging studies have all had their own shortcomings with frequent or occasional false positive and negative results [1]. On the other hand, some don't feel obliged to operate on every patient based on imaging diagnosis of malrotation [2]. The number of radiologic publications addressing the diagnostic difficulties regarding malrotation is steadily increasing despite an earlier publication in which UGI was introduced to be the diagnostic modality of choice [3] The mere fact that SPR has allocated a 90 minute panel discussion in its annual meeting is the testimony to the frustration of the membership as they attempt to correctly exclude or preclude malrotation in their daily practice.

Barium enema

Practically everyone now agrees that neither a normally positioned cecum in RLQ excludes malrotation nor a high positioned cecum in RUQ precludes it. However, if the cecum is clearly in LUQ or LLQ, diagnosis of malrotation must be considered with exception of situs inversus [4].

UGI

The most important landmark used in UGI is the position of duodenojejunal junction (DJJ) at the level of left L1 pedicle, the level at which often either pylorus or the duodenal bulb (DB) are situated [5, 6]. It is assumed that the DJJ is anchored at that position by the ligament of Trietz (LT). This is only true if the duodenal sweep is c-shaped and the following factors don't interfere with the position of DJJ.

1. Dilated stomach and small bowel loops [6].
2. Positive left retroperitoneal force pushing the DJJ to the contra lateral side [7].
3. Left intraperitoneal mass effect, transplanted left lobe of the liver, etc. [8]
4. Negative force in right hemi abdomen, pulling the DJJ rightward (personal observation).
5. Redundant duodenum that makes the recognition of the exact point of the DJJ is difficult, leaving an element of guess and uncertainty in the radiologist's interpretation [6].
6. The most frequent and the least recognized of all the factors is the right sided position of the proximal jejunum, pulling the DJJ rightward.

Some authors have pointed to the normal variations of the position of DJJ in transverse and cephalocaudad directions, indicating that the DJJ need not be exactly at the level of left L1 pedicle to be normal [9]. Furthermore, it is not clear how strongly the LT can secure the DJJ position. Manual studies demonstrate mobility of DJJ in infants and young children [9–11]. In older children mobility is less or absent but not necessarily due to firm attachment of DJJ [9]. It is more difficult to displace DJJ manually in older children because the resistance of abdominal wall muscles and guarding in these patients [9]. Displacement of DJJ due to retroperitoneal mass indicates that the DJJ is not so firmly attached to where it is expected to be.

Some authors indicate that in lateral projection of an UGI, the 2nd and 4th portions of duodenum (D2 and D4) are posterior and run more or less parallel to each other [6]. This also has its own shortcomings because there is no landmark in lateral projection to indicate that D2 and D4 are posterior enough to be in retroperitoneal space. In fact, the anteroposterior distance between an intraperitoneal D3 (malrotation) and the retroperitoneal D3 (normal) is only the thickness of the peritoneal reflection plus the anteroposterior dimension of the SMA. This only amounts to a total of 2–3 mm which is not recognizable in a lateral projection of an UGI because of lack of a normal anatomic point of reference. Furthermore, in practice, unparallel orientation of D2 and D4 can be seen normally and, on the other hand, D2 & D4 can be posterior in lateral view, despite an obvious malrotation in AP view.

The deviation from the norms and false positive studies, in our experience, is due to rightward position of proximal jejunum that distorts the position of D4 and DJJ as the pulling force of the rightward jejunum overcomes the anchoring strength of the LT. This phenomenon explains why the configuration of the duodenal sweep and position of the DJJ varies from one study to another in two different studies and in two different times in the same patient.

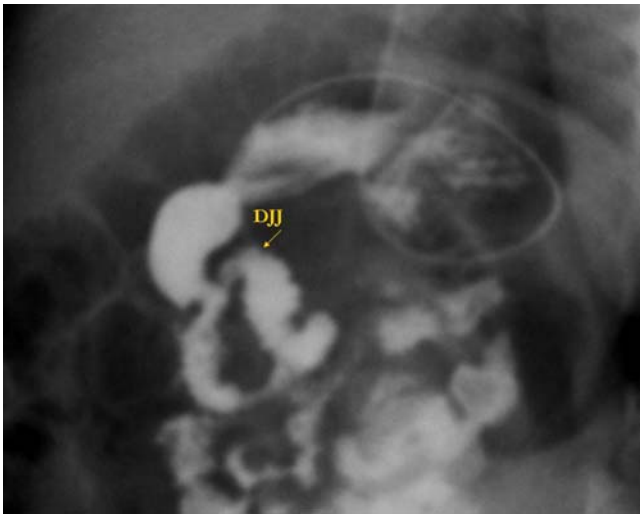

Fig. 1 UGI with unusual location of duodenojejunal junction (*DJJ*) or flexure. Patient is slightly rotated. The proximal jejunal loops are on the right side

The most frightening shortcoming of the UGI is when the patient has only a non occlusive 180° volvulus, flipping the DJJ from right to left and displaying the DJJ in a non-incriminating position without creating a transition at the point of the twist. This can result in a false-negative study [10]. This false-negative aspect of UGI may be coupled by the false-negative clinical input of "nonbilious vomiting" which can occur in about 14% of patients with midgut volvulus [12].

The rate of false positive and negative UGI for malrotation varies from one series to another and is observer dependent. Irrespective of the rate, the above mentioned shortcomings are encountered with such annoying frequency that can result in misleading findings in UGI, with an element of guessing and uncertainty in its interpretation.

Ultrasound

Initially, it was assumed that if SMV is not to the right of SMA in axial plain, at the level of the junction of SMV with the portal vein, the findings will be highly sensitive for malrotation [13]. The diagnostic sensitivity drops if SMV is anterior to SMA [14, 15]. If the SMV is clearly to the right of SMA, it is believed to exclude malrotation [13]. Our experience in some patients indicates otherwise. Some have suggested that if SMA is truncated in longitudinal imaging of the abdomen and if it is not parallel with and anterior to the abdominal aorta, this should signal malrotation-volvulus [15]. However, the truncated SMA merely indicates that SMA is not midline. The position of SMA, in our experience, is more dictated by the position of the proximal jejunal loops and malposition of jejunum is common and need not indicate malrotation.

Others consider the "whirlpool sign" highly sensitive for diagnosis of malrotation by US and refer to this pattern as clockwise or counterclockwise interchangeably [17–21]. Yet some investigators point to the shortcomings of this sign as well [22]. Most authors believe that ultrasound cannot exclude malrotation [23], echoed by the commentary of one of the leading scholars of the field of pediatric radiology [23], reinforcing the current belief that UGI is the modality of choice and the "reference standard" in diagnosis of malrotation.

CT

The ultrasonographers' initial contribution stemmed from the initial CT publications regarding the relative positions of SMA and SMV [24–27]. Later the swirling whirlpool sign was introduced—first as a reliable and later as a non-reliable sign for malrotation-volvulus [22]. The authors of both CT and US publications have consistently misidentified the nature of the swirling vessel to represent the SMV. However, our review of 150 contrast-enhanced CT proves the swirling vessel to be a branch of jejunal vein often the 1st jejunal vein, not the SMV itself, that circles around clockwise and runs between the SMA and aorta before joining the SMV. Three findings were consistently observed in our cases with clockwise whirlpool sign (clockwise indicating the direction of blood flow):

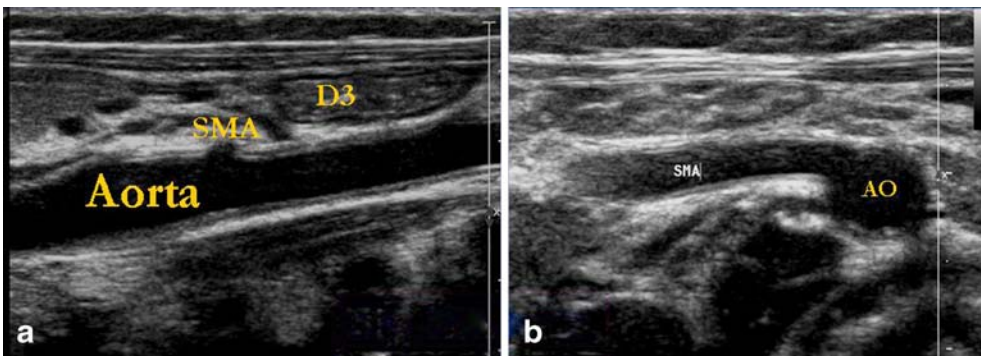

Fig. 2 **a** Longitudinal image of aorta showing truncated SMA. The D3 is anterior to the aorta but not behind the SMA in this plain because the SMA is not aligned with the aorta due to right sided jejunum and is horizontally oriented in transverse plain as shown in **b**

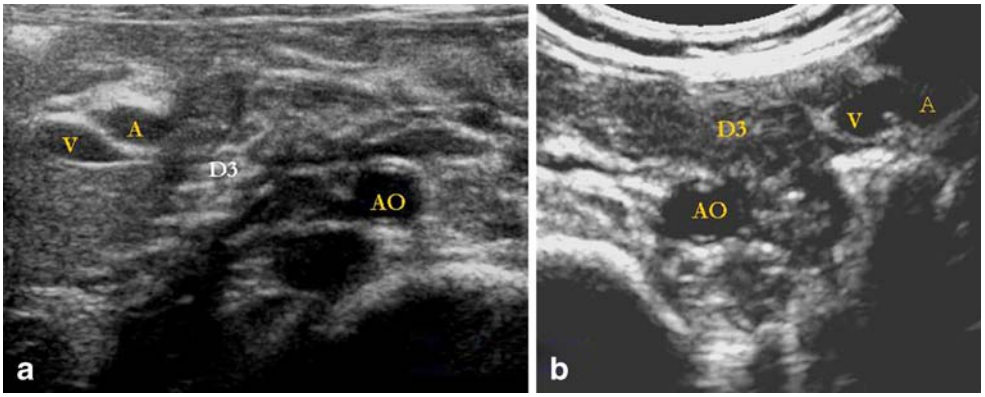

Fig. 3 (**a**) Transverse imaging showing the SMA (*A*) and SMV (*V*) way to the right of aorta because of rightward jejunum with *D3* in between. (**b**) After manual compression on the right side, the mesenteric vessels are moved to the left of aorta (*V*&*A*) with D3 in between AO and mesenteric vessels

A. In all the patients with the whirlpool sign in axial plain, the SMA and SMV both had a vertical course, in cephalocaudal direction in coronal & sagittal reconstruction. If the swirling vein, as claimed by previous authors, were to represent the SMV, then how could the SMV be swirling with a horizontal course in axial plain and have a vertical orientation in coronal and sagittal plains?
B. In every case with clockwise whirlpool sign, the proximal jejunum was to the right of the midline folding the jejunal vein from its expected leftward position to right hemi abdomen, creating the whirlpool sign. As the jejunum folded to the right so did the jejunal vein. In most normal subjects, the jejunal vein (s) runs between the SMA and aorta. The rightward position of the proximal jejunum can pull the DJJ rightward and away from its expected left L1 pedicle position. This displaces the D4 anteriorly and in doing so, the D4 won't be parallel with D2 in lateral projection, causing false positive interpretation of UGI.

In few patients with previous or follow-up CT study, the jejunal loops were displaced to the right when the whirlpool sign was positive and relocated on the left side when whirlpool sign was not seen.

C. In every patient with whirlpool sign, the D3 was between SMA and AO.

We believe that the shortcomings of current imaging techniques are all due to not considering the following facts:

1. Intraoperatively, in malrotation, the 3rd portion of duodenum (D3) is always anterior to the SMA (intraperitoneal), not posterior to the SMA (retroperitoneal).
2. In normal individuals, irrespective of age, the D3 is always situated between SMA & AO.
3. Retromesenteric D3 indicates that the gut has reached its final embryologic destination fixed in retroperitoneal compartment, as it should be, hence excluding malrotation.
4. Excluding malrotation always excludes midgut volvulus (MGV)
5. Retromesenteric D3 is a constantly demonstrable and 100% trustworthy landmark, whereas, the ligament of Treitz is invisible and is only a presumptive structure for the imagers. One can only infer to the existence of LT by

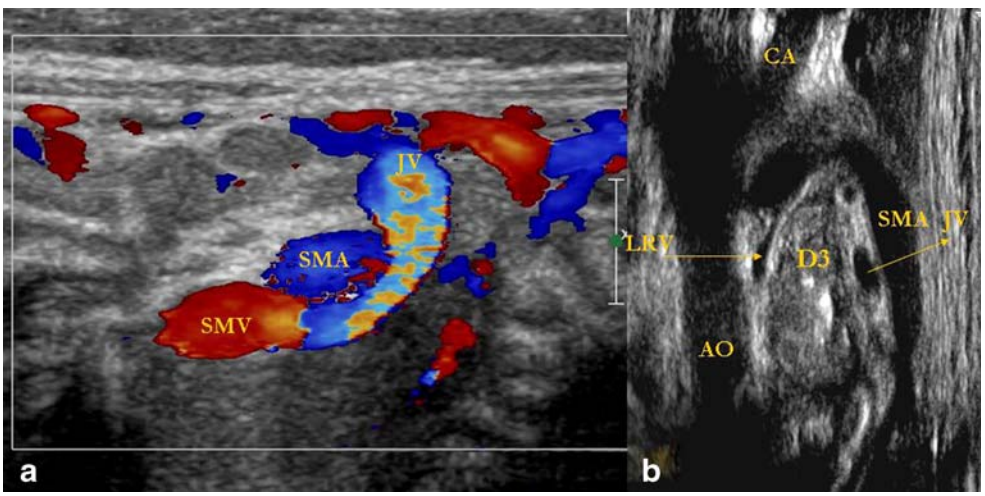

Fig. 4 (**a**) Clockwise swirling of the rightward displaced jejunal vein, circling around the SMA and emptying into the SMV. (**b**) After compression on the right side, the SMA is aligned with the AO with D3 in between in sagittal plain. Note the left renal vein and jejunal veins (*JV*) also behind the SMA and anterior to the aorta

demonstrating the retromesenteric D3 not by a "normal" position of DJJ.
6. DJJ can be displaced by many factors, but D3 is fixed irrespective of these factors.

In our series and as reported in CT literature [28], retromesenteric D3 was always recognizable in normal individuals with or without "whirlpool sign."

Considering these facts, we offer ultrasound as an alternative to CT to illustrate retromesenteric D3 in axial and sagittal plains. This in one hand is the imaging modality of choice and the most trustworthy one and, in another hand, is in keeping with the ALARA principle and our campaign motto of "Image Gently."

Ultrasound technique

Using a high resolution linear transducer and gentle grading compression, serial axial images will be obtained in cephalocaudad direction and the following structures will be displayed.

1. Confluence of splenic vein, SMV and the portal vein with the pancreas usually in the image.
2. At lower level, the left renal vein will be seen crossing from left to right between the SMA and the AO.
3. Slightly lower the left sided jejunal vein will be visualized, often running between SMA and AO before joining the SMV.
4. If jejunum is on the right side, the whirlpool sign with clockwise flow direction will be seen.
5. Below the left renal vein and the jejunal vein the D3 with mucosa and muscularis will be demonstrated between SMA and AO, running from right to left across the spine.
6. The only loop of bowel that runs between SMA and AO (retroperitoneal) is D3; the jejunum is intraperitoneal.
7. If D3 is not visualized due to technical factors, by demonstrating parallel course of SMA and SMV in coronal plain or vertical orientation of SMA & SMV in sagittal plain, midgut volvulus will be excluded because both will be twisted with volvulus.
8. If SMA is not anterior to AO with D3 in between, with manual compression on the right side of the abdomen, the rightward jejunum can be pushed to the left side and the SMA will be aligned with the AO with D3 in between.

Conclusions

We conclude that the position of DJJ is the wrong horse to bet on in malrotation work-up. Why chasing a variable when we can bank on a constant that indicates that the gut has reached its final embryologic destination and the transverse duodenum (D3) is secured, as it should be, in retroperitoneal compartment, immune of undergoing volvulus.

Recommended imaging philosophy to diagnose or exclude malrotation

1. Demonstrating a retromesenteric D3 should be attempted in every child undergoing an ultrasound study. For the rest of his/her life, that person will not have malrotation and will not develop MGV.
2. Retromesenteric D3 obviates UGI.
3. Premature babies prone to develop NEC, sepsis, etc. not infrequently have bilious vomiting. In the same setting of initial head ultrasound survey, malrotation can be and should be excluded obviating the need for an UGI in the later date when the baby has bilious or nonbilious vomiting.
4. Parents of children with normal position of D3 should be informed that their child need not be studied for malrotation in case of vomiting which is a very common childhood symptom.
5. If a retromesenteric D3 is visualized, it matters not where the position of DJJ is, whether the duodenal sweep is C-shaped, Z-shaped, redundant or not, or what are the relative positions of mesenteric vessels or any other currently used criteria for diagnosis or exclusion of malrotation.
6. Normal position of D3 in US is therefore the "reference standard" not the "normal" position of DJJ in UGI.
7. UGI does not prove that the D3 is retroperitoneal and runs behind the SMA, US does.

A representative case exemplifies the shortcomings of UGI and currently used US criteria for diagnosing malrotation (Figs. 1, 2, 3, 4).

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