

The efficiency of sonography in diagnosing volvulus in neonates with suspected intestinal malrotation

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Abstract

This study is to prospectively evaluate the efficiency of sonography for volvulus diagnosis in neonates with clinically suspected intestinal malrotation.

A total of 83 patients with suspected intestinal malrotation who underwent detailed abdominal sonography and upper gastrointestinal contrast study were included. Malrotation was characterized by inversion of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) in sonographic examination. The “whirlpool sign” of Color Doppler Sonography was recognized as a characteristic for malrotation with volvulus. The degrees of rotation of the SMV winding around SMA were also detected by sonography. Surgery was performed in patients with sonography diagnosed malrotation.

A total of 39 patients were sonographically diagnosed as malrotation which was subsequently confirmed by surgery. The sensitivity and positive predictive value of the sonographic diagnosis were both 100% (39/39). The sensitivity, specificity and accuracy of “whirlpool sign” for the detection of midgut volvulus were 95.2% (20/21), 88.9% (16/18), and 92.3% (36/39), respectively. Greater degrees of rotation (equal or greater than 720°) showed higher risk (odds ratio, 5.0; $P < .01$) for intestinal necrosis occurrence.

Sonography is more accurate in diagnosing suspected malrotation than upper gastrointestinal contrast study. Specific sonographic “whirlpool sign” related to volvulus may be used as a potential indicator for intestinal necrosis. In addition, sonography can exclude malrotation and may help the diagnosis of other diseases, such as annular pancreas and duodenal atresia.

Abbreviations: CT = computed tomography, DU = duodenal, GI = gastrointestinal imaging, NPV = negative predictive value, PPV = positive predictive value, SMA = superior mesenteric artery, SMV = superior mesenteric vein, STO = stomach.

Keywords: malrotation, midgut volvulus, neonate, sonography, upper gastrointestinal, whirlpool sign

1. Introduction

Intestinal malrotation is one of the most common embryonic malformations in the digestive tract. Malrotation with volvulus is a potentially life-threatening condition.^[1] Sudden onset of bilious vomiting with a flat abdomen in the neonatal period strongly suggests the diagnosis of malrotation. One of the consequences of malrotation is midgut volvulus. Most neonates with midgut volvulus usually present this complication within 7 days after birth, and up to 80% of patients with midgut volvulus develop this complication within the first month of life.^[2]

Early diagnosis of malrotation is critical as delayed diagnosis can lead to necrosis of the midgut and mortality.^[3–5] Diagnosis of malrotation mainly relies on gastrointestinal imaging (GI) in the past

but the radiologic results are not always reliable.^[6] Sonography is more valuable for accurate diagnosis of malrotation than an upper gastrointestinal contrast study.^[7] Currently, sonography is considered as the preferential tool for the evaluation of suspected malrotation in children.^[8–10] Malrotation could be diagnosed by sonography, in which inverse orientations of the superior mesenteric artery (SMA) and the superior mesenteric vein (SMV) are featured signs of malrotation. In addition, “whirlpool sign” is an imaging characteristic of midgut volvulus and has a high predicting value for volvulus.^[8–10] Malrotation or malrotation with volvulus can be effectively diagnosed based on these characteristics.^[8–10] Ultrasound diagnosis has the advantage of no radiation exposure. The rapid and accurate diagnosis of volvulus^[11] by ultrasound helps to establish the diagnosis of malrotation in time and thus allows for urgent surgical intervention to avoid bowel necrosis. However, not all cases of malrotation have abnormal SMV/SMA orientations on ultrasound images.^[12] On the one hand, malrotation can occur when the mesenteric vessels are rotated correctly.^[1,12,13] On the other hand, not all cases of “whirlpool sign” were found to suffer from malrotation.^[7] The aim of this study is to evaluate the potential value of sonography in identifying or ruling out malrotation or volvulus in neonates, and to predict intestinal necrosis by degrees of rotation of the superior mesenteric vessels.

2. Materials and methods

2.1. Patients

This prospective study recruited 83 infants (male: 45; female: 38) aging from 1 day to 31 days with clinically suspected malrotation in the Shandong Qianfoshan Hospital between February 2013

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and July 2016. All patients presented with acute clinical symptoms suggestive of malrotation, such as irritability, poor feeding, and bilious vomiting. Malrotation was diagnosed immediately after scanning by a pediatric sonographer, who was blinded to other examination findings. Patients who had a history of abdominal surgery or pyloric muscle hypertrophies were ruled out. The research was approved by the Hospital Institutional Clinical Research Ethics Committee. All informed parental consent forms were signed.

2.2. Ultrasonographic examination and the upper gastrointestinal contrast study

The sonographic examinations were performed for all recruited infants by a pediatric sonographer with 3 years of pediatric sonography experience. He used a Philips iU22 scanner (Philips Medical Corporation, Hudson, WI), a C5-1-MHz curvilinear transducer (Philips Medical Corporation) or a L12-5-MHz linear array transducer (Philips Medical Corporation). Briefly, for clear visualization, 50 mL of water were instilled via the nasogastric tube before sonographic examination. During water instillation, the patients were placed in the right lateral decubitus position to allow water flowing into the duodenum as soon as possible. Then, the patients were placed in the supine posture for ultrasonographic examination. The antropyloric and duodenal portion were scanned longitudinally first, followed by the descending portion of the duodenum lateral on transverse scanning below the pancreatic head and more distal intestine. The position of superior mesenteric vessels, collapse of bowel loops, and other abnormalities were recorded. Color Doppler imaging was used to verify the SMA and SMV, and whether there was a “whirlpool sign.” Malrotation was recorded if an inverse orientation of the SMA and SMV was shown. Malrotation and midgut volvulus were ruled out if none of the above features was found. The diagnosis of malrotation or midgut volvulus was confirmed by surgical exploration. Intestinal necrosis was also predicted by the degrees of clockwise rotation with the SMV winding around SMA. Finally, the rest of the abdomen was scanned to exclude other abnormalities.

The upper gastrointestinal contrast study was performed for all recruited infants by another radiological specialist with 3 years of experience in pediatric abdominal radiology. The malrotation was indicated by duodenojejunal flexure at the right of the vertebral bodies on UGI images.

2.3. Other imaging examinations

For infants who did not receive surgery, other imaging examinations (such as barium enema, or computed tomography (CT)) were performed. CT examination was applied to critically ill patients. Malrotation was excluded if no sign indicating malrotation was shown by these examinations.

2.4. Statistical analysis

SPSS software (version 13.0) was used for all data analysis. Data were analyzed using Chi-square test. A P -value $< .05$ was considered statistically significant.

3. Results

3.1. Diagnostic results of malrotation

Of the 83 infants, 39 had malrotation as diagnosed by sonographic examination, and the results were subsequently confirmed by

Table 1

Final diagnoses as confirmed by surgery and other imaging studies in 83 patients.

| Diagnosis | n = 83 |
|---|--------|
| Surgery | |
| Malrotation | 39 |
| Annular pancreas | 2 |
| Duodenal atresia | 1 |
| Duodenal stenosis | 2 |
| Descending duodenal web | 1 |
| Other imaging findings (upper gastrointestinal study, barium enema, or computed tomography) | |
| Nonmalrotation | 38 |

surgery. Six infants with malrotation were eventually excluded due to the sonography and/or surgery demonstrated abnormalities, including annular pancreas (2 cases), duodenal atresia (1 case), duodenal stenosis (2 cases), and descending duodenal web (1 case). The remaining 38 infants were finally diagnosed with non-malrotation by barium enema, or CT examination (Table 1).

The results of sonographic examination, upper gastrointestinal barium, and surgery were compared. The results are shown in Table 2. The ultrasonographic characteristics of inversion of the SMA and SMV identified all of the 39 patients with malrotation that were later surgically confirmed, with a sensitivity of 100%. As shown in Fig. 1, SMV (thick arrow) appeared to the left side of the SMA (thin arrow). In contrast, the positive upper gastrointestinal contrast study only identified 36 out of the 39 patients with malrotation, with a sensitivity of 92.3%. The duodenojejunal flexures were not shown clearly because the obstruction blocked the contrast agent from entering across the horizontal part of duodenum or the descending duodenum in the remaining 3 infants. The negative predictive value (NPV) of upper gastrointestinal barium diagnosis was 93.6% (44/47). Nevertheless, the positive predictive value (PPV) and specificity of both sonographic diagnosis and upper gastrointestinal barium diagnosis were 100% (36/36) and 100% (44/44), respectively.

3.2. Diagnostic results of volvulus

We investigated the accuracy of sonographic diagnosis for volvulus in the 39 patients with malrotation. Of them, 21 cases were complicated by volvulus as confirmed by surgery. “Whirlpool sign” of sonography was observed in 22 cases (Table 3) corresponding to a clockwise wrapping of the SMV around the SMA. The SMV (thick arrow) can be seen encircling SMA (thin arrow; Fig. 2A), and sonogram showed the

Table 2

Comparison of sonography, upper gastrointestinal barium, and surgery results for the diagnosis of the 83 neonates with suspected malrotation.

| Malrotation examinations | Surgical findings | |
|--|-------------------|-------------------|
| | Positive (n = 39) | Negative (n = 44) |
| Sonographic diagnosis (inversion of SMV/SMA) | | |
| Positive (n = 39) | 39 | 0 |
| Negative (n = 44) | 0 | 44 |
| Upper gastrointestinal barium diagnosis | | |
| Positive (n = 36) | 36 | 0 |
| Negative (n = 47) | 3 | 44 |

SMA = superior mesenteric artery, SMV = superior mesenteric vein.

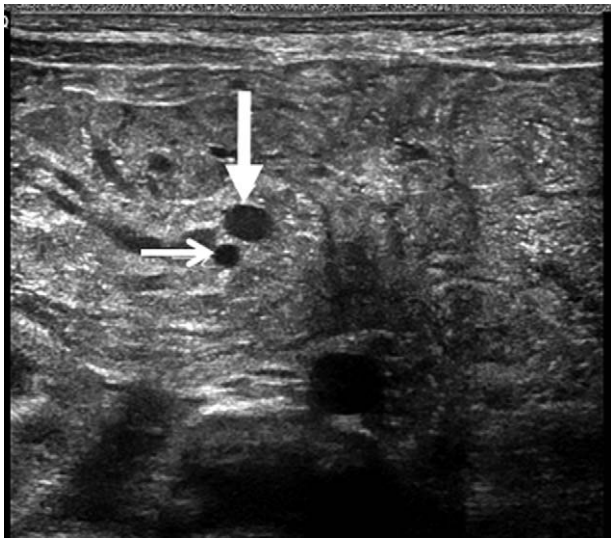


Figure 1. Ultrasound transverse section image in a 5-day-old boy with bilious vomiting. It is shown that SMV (thick arrow) is visible to the left side of the SMA (thin arrow).

“whirlpool sign” with the SMV (thick arrow) surrounding the SMA (thin arrow; Fig. 2B). The dilation of the stomach (STO) and the duodenum (DU) was also observed (Fig. 2C). Two false-positive and 1 false-negative volvulus were detected by “whirlpool sign.” For the 2 false-positive cases, the sign was caused by upper jejunum diverticulum in 1 case, and in the other child a postoperative intussusception was noted, a 0.2 cm × 0.3 cm × 0.5 cm lymphangioma acting as a lead point for the intussusceptions. The false-negative case received CT examination and was subsequently confirmed by surgery. Overall, “whirlpool sign” of sonography for the diagnosis of volvulus in patients with malrotation had the sensitivity, specificity, PPV, NPV, and accuracy of 95.2% (20/21), 88.9% (16/18), 90.9% (20/22), 94.1% (16/17), 92.3% (36/39), respectively (Table 3). Additionally, the false-positive rate (9.1%; 2/22) and false-negative rate (5.9%; 1/17) were low.

3.3. Sonographic diagnosis number of twists of malrotation and intestinal necrosis

Sonography also allowed an accurate estimation for the number of twists of the SMV around SMA, which ranged from 270 to 1080° (Table 4). Degrees of rotation by sonographic diagnosis

Table 3
Whirlpool sign of sonography with or without volvulus in neonates with malrotation (n=39).

| Whirlpool sign | Volvulus/nonvolvulus [*] (n=21/18) | Sensitivity % (n) | Specificity % (n) | PPV % (n) | NPV % (n) | Accuracy % (n) |
|-----------------|---|-------------------|-------------------|---------------|---------------|----------------|
| Positive (n=22) | 20/2 | 95.2% (20/21) | 88.9% (16/18) | 90.9% (20/22) | 94.1% (16/17) | 92.3% (36/39) |
| Negative (n=17) | 1/16 | | | | | |

NPV = negative predictive value, PPV = positive predictive value.
* Confirmed by surgery.

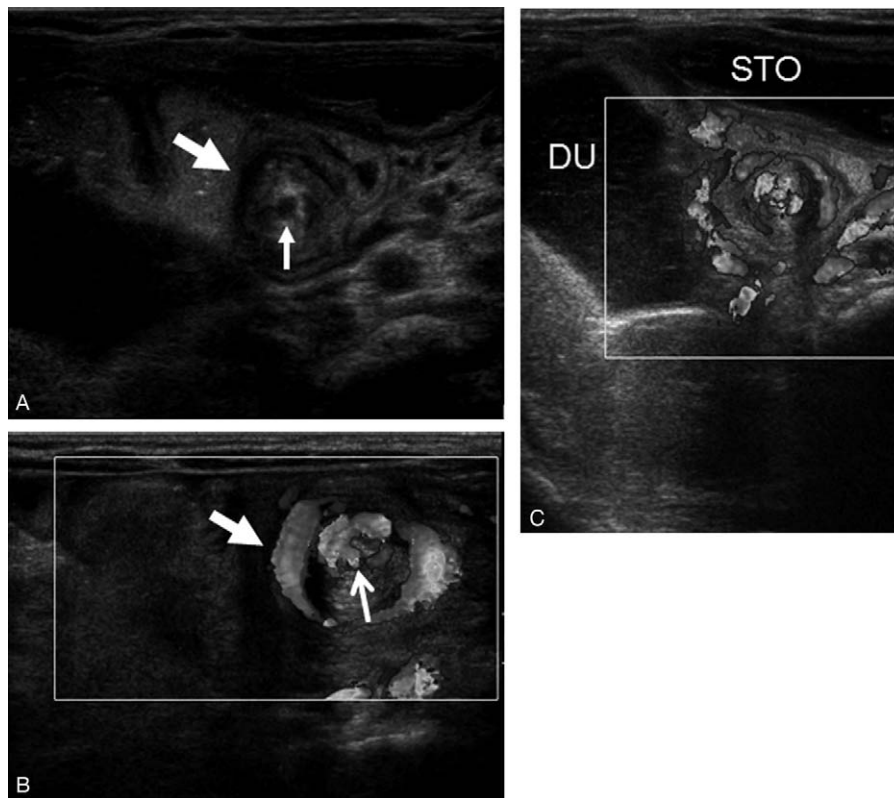


Figure 2. Whirlpool sign: transverse ultrasound of mesenteric root in a 12-day-old boy with bilious vomiting and a dilated abdomen. (A) The SMV (thick arrow) can be seen encircling the SMA (thin arrow). (B) Color Doppler ultrasound image of the whirlpool sign. Whirlpool sign with complete wrapping of the SMV (thick arrow) around the SMA (thin arrow) in clockwise rotation of the mesentery. (C) Dilation of stomach (STO) and duodenum (DU) in intestinal malrotation with volvulus.

Table 4**Diagnosis for the degrees of rotation by ultrasound in neonates with whirlpool sign.**

| Number | Degrees of rotation, ° | Sonographic diagnosis cases (n=22) | Surgically proved cases (n=21) | Accuracy, % |
|--------|------------------------|------------------------------------|--------------------------------|-------------|
| 1 | 270 | 0 | 1 | 0 |
| 2 | 360 | 10 | 10 | 100 |
| 3 | 540 | 5 | 4 | 80 |
| 4 | 720 | 6 | 5 | 83 |
| 5 | 1080 | 1 | 1 | 100 |

Table 5**Correlation of intestinal necrosis and degrees of rotation in 21 neonates with volvulus.**

| Degrees of rotation | Intestinal necrosis | | Total | Incidence, % |
|---------------------|---------------------|----|-------|--------------|
| | Yes | No | | |
| ≥720° (n=6) | 2 | 4 | 6 | 33 |
| <720° (n=16) | 1 | 14 | 15 | 6.7 |
| Total | 3 | 18 | 21 | 14 |

have a high accuracy, and 360° was the most commonly detected twist (the diagnostic rate was 100%). Although only 1 case twisted 1080°, sonography can also accurately diagnose patients with 1080° rotation (Table 4). However, sonography was unable to accurately diagnose the patient with 270° twist.

We found that 21 infants with malrotation had volvulus confirmed by operation, among them 6 patients who had rotations equal or greater than 720° and 15 patients had rotations less than 720° (Table 5). Incidence of intestinal necrosis in patients with different rotation degrees is also shown in Table 5. Incidence of intestinal necrosis in cases with rotations equal or greater than 720° was 33.3% (2/6), whereas the incidence in cases with rotations less than 720° was 6.7% (1/15). However, Chi-square test confirmed no statistically significant relationship between the degrees of rotation and intestinal necrosis.

4. Discussion

Malrotation is a congenital abnormality that induces upper gastrointestinal obstruction in neonates with intestinal malrotation and it requires emergent operations to prevent the catastrophic complication of volvulus and bowel necrosis.^[14] Thus, early diagnosis of intestinal malrotation is of great importance. In the past, upper gastrointestinal examination was the preferred examination for the diagnosis of intestinal malrotation, although it had the drawbacks of low sensitivity, radiation exposure, and difficult positioning.^[9] In addition, Barium enema is no longer a routinely used diagnostic method for malrotation due to the fact that approximately 20% of children with malrotation have a normally positioned cecum.^[15]

In contrast, ultrasonography has potential benefits of portability, low levels of radiation exposure, and applicability to critically ill children who might be too sick to take a gastrointestinal contrast study. Therefore, ultrasonography plays an important role for neonatal care in the intensive care unit. In recent years, ultrasonography has been introduced as an alternative method for the diagnosis of malrotation, with an emphasis on the relationship of the SMV and SMA and the so-called “whirlpool sign” in cases of volvulus. Studies have reported that sonographic findings of abnormal relative positions of the SMA and SMV have high sensitivity in diagnosing intestinal malrotation.^[16,17] Inversion of the superior mesenteric

vessels confirms the diagnosis of malrotation. Studies have suggested that the inversion of the SMV (i.e., the SMV to the left of the SMA) is found in 100% malrotation cases.^[1,5] In this study, all of the diagnoses were confirmed surgically to have a left-sided SMV. The main advantages of ultrasound are noninvasiveness and simplicity.^[13] Alehossein et al^[9] reported that sonographic accuracy eliminated the need for further diagnostic tests. In this study, 6 patients with other diseases were also successfully detected by ultrasonography.

The main features of malrotation on an upper gastrointestinal study include duodenojejunal flexure at the right of the midline. In this study, the upper gastrointestinal contrast study failed to identify malrotation in 3 infants. This failure may have occurred because the contrast agent did not pass the horizontal part or the descending part of duodenum due to obstruction.

Sonographical “whirlpool sign” is sensitive in detecting volvulus,^[16] with the reported sensitivity of 86%, specificity of 92% and PPV of 89%.^[8] The specificity and the NPV of “whirlpool sign” in midgut volvulus detection were reported as 99% and 97.1%, respectively.^[10] Therefore “whirlpool sign” of sonography is considered as a reliable indicator for the presence of volvulus. Consistently, our study showed that the “whirlpool sign” could detect volvulus with the sensitivity, specificity and accuracy of 95.2%, 88.9%, and 92.3% respectively. Our results also showed that this detection method had a high NPV (94.1%). Importantly, midgut volvulus might cause intestinal necrosis, which could be used as an indication for surgical intervention.^[5,18,19]

It should be noted that not all of “whirlpool sign” can be considered as cases of volvulus^[8,14] and other causes can contribute to “whirlpool sign”^[20]; meanwhile, absence of a “whirlpool sign” cannot reliably exclude volvulus either. In this study, a whirlpool sign was observed in 2 patients who were not volvulus cases as excluded by surgery. Besides, although Karmazyn et al^[21] suggested that a normal sonographic finding definitively ruled out the danger of midgut volvulus, we have found that 1 patient with normal ultrasound was actually suffering from midgut volvulus as confirmed by CT scanning and surgery. Sometimes, further radiographic contrast studies are required as a supplement to ultrasound diagnosis.^[9,22]

Ultrasonography can also be used as an accurate estimation on the number of twists of the SMV. However, sonography was

unable to accurately diagnose the twist with rotation less than 270°. This may be because twists with rotation degrees lower than 270° are hard to detect and the experience of the examiner also may play a role. We also found that the twists of 360° were the most frequently observed twist in the 21 patients, and the patients with twists equal or more than 720° tended to have intestinal necrosis (33.3%; 2/6). In contrast, of the 15 patients with twists less than 720°, only one intestinal necrosis was identified (incidence: 6.7%). However, there was no statistically significant relationship between the degrees of rotation and intestinal necrosis. We speculate that this is caused by a limited number of patients is and future studies with larger sample size are needed. Our results indicate that sonography can accurately diagnose degrees of rotation to fulfill the need of surgical intervention and avoid intestinal necrosis.

The main limitations of this study include that the follow-up period is short and the sample size is small. Thus, we cannot conclude that all negative sonographic findings and upper gastrointestinal contrast findings are true-negative results. In many patients malrotation is not diagnosed until adulthood,^[14,23] in some patients false-negative sonographic results may have occurred.

5. Conclusions

In conclusion, our study shows that sonography may be more valuable for the accurate diagnosis of suspected malrotation (inversion of the SMA and SMV) than upper gastrointestinal contrast studies. The upper gastrointestinal contrast studies have low diagnostic accuracy and a slightly higher false-negative rate. The specific sonographic “whirlpool sign” can be used to evaluate degrees of rotation and moreover, sonography can exclude malrotation and provide additional diagnostic information, such as annular pancreas and duodenal atresia, etc.

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