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The Diagnostic Value of Mesenteric Vessel Abnormalities on Ultrasound for Malrotation

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Abstract

Background: Malrotation is an emergency, mainly in pediatric cases, and needs a timely and prompt diagnosis. Upper gastrointestinal (GI) contrast study used to be an acceptable modality in the diagnosis of malrotation; however, it has many disadvantages. In this regard, sonography has tried to take the place of upper GI studies. The aim of our study is to assess sonography and upper GI series as diagnostic methods for malrotation.

Methods: In a cross-sectional study, 154 pediatric cases suspected of malrotation were enrolled. The patients underwent upper GI series and sonography. In the case of sonography, two different findings, including inversion of the superior mesenteric vein (SMV) and superior mesenteric artery (SMA) and deviation (abnormal pathway) of the mesenteric artery were assessed. The sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) of different sonography findings and upper GI study were calculated.

Result: Different sonography findings showed various diagnostic values. Inversion of SMV/SMA had a sensitivity of 58.87%, specificity of 36.17%, PPV of 67.74%, and NPV of 27.86%. Furthermore, the sensitivity, specificity, PPV, and NPV of deviation (abnormal pathway) of mesenteric vessels were 89.87%, 17.02%, 64.54%, and 50%, respectively. Taking into consideration the two findings together, sonography showed a sensitivity of 73.83% and PPV of 67.74%. Moreover, the sensitivity, specificity, PPV, and NPV of upper GI studies were 82.5%, 100%, 100%, and 50%, respectively.

Conclusion: Mesenteric vessel abnormalities may be valuable in detecting malrotation, but still upper GI contrast study is better. Development of other sonographic markers of malrotation, especially for different ages, is necessary.

Key Words: Malrotation, Mesenteric vessels abnormalities, Ultrasound.

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1- INTRODUCTION

Malrotation is a congenital anomaly caused due to the abnormal rotation of the embryonic gut. The condition is believed to be a problem of infancy and only 10% of the cases happen after one-year-old age (1).

The true incidence of the condition is not fully known, as it may occur in an asymptomatic manner. The disorder may be diagnosed as an incidental imaging finding through upper gastrointestinal (GI) contrast studies in 2 out of 1000 people, known as nonrotation. However, the incidence of symptomatic Malrotation is 1 in 6000 cases of live births (2).

The condition may present itself with a variety of symptoms. One of the most prevalent symptoms is bilious vomiting. Moreover, the patients may present with symptoms of abdominal pain, distention, peritonitis, hematochezia, or even hemodynamic instability (3).

Rarely, the condition may develop chronic diarrhea, mal-absorption, chylous ascites, pancreatitis, and failure to thrive (2, 4). A recently published article during a retrospective study for a fifteen-year period revealed that 68% of the patients had vomiting and 57% had abdominal pain (5).

In fact, malrotation may result in volvulus, which is associated with catastrophic results such as intestinal ischemia and mid-gut gangrene, or even mortality. With this regard and considering the emergency pattern of this disorder, timely and prompt diagnosis is very important for a good surgical outcome (6, 7). However, the use of diagnostic methods is really controversial (8). Some believe in using upper gastrointestinal series in the diagnosis of Malrotation (9-11). However, the radiation, difficulty in patient's positioning, and physical traumas such as the risk of hypothermia have limited the applicability of this method. Moreover, it seems that the modality is not accurate enough (8, 9). Then, sonography is usually considered as the first-choice method in these patients (12-14).

However, the modality is usually operator-dependent and thus it has a varying accuracy. In fact, it is reported that the normal retroperitoneal position of the third segment of the duodenum is the best sonography marker for malrotation, however, it is not easy to identify and only very expert sonographists can find it. Different sonography findings such as, inversion of the superior mesenteric artery and superior mesenteric vein, and deviation (abnormal pathway) of mesenteric vessels are, still, assessed for the diagnosis of this disorder. Unfortunately, mesenteric vessel abnormalities are not exclusive to the diagnosis of malrotation and they are also seen in bowel obstruction, diaphragmatic hernia, and space occupying lesions (8, 13, 14).

Here, we investigated the diagnostic value of different sonography findings for detecting malrotation.

2- MATERIALS AND METHODS

2-1. Study design and population

This cross-sectional study was conducted on children with suspected intestinal malrotation in the Surgery Department of Akbar Children's Hospital affiliated to Mashhad University of Medical Sciences, Mashhad, Iran, between 2017 and 2021.

2-2. Inclusion and exclusion criteria

The inclusion criteria encompassed any child who was suspected of malrotation based on clinical findings including signs and symptoms, such as irritability, poor feeding, and bilious vomiting. Patients who had a history of abdominal surgery or pyloric muscle hypertrophies or previous abdominal surgery were ruled out.

2-3. Imaging studies

As ultrasound was the first diagnostic method used in the examination of abdominal complaints at our hospital, the sonographic examinations were conducted for all included children by two experienced pediatric radiologists with 10 years of experience. Gray-scale sonography was performed with sonographic machines such as GE (voluson, E6) and MyLab class C (Esaote, Italy) incorporating 7.5-12 MHz linear and 2.5-5 MHz curved probes.

For detecting the direction and location of mesenteric vessels, the individuals were scanned in the supine position with the transducer positioned in the midline of the anterior upper abdomen. For a better visualization of the mesenteric vessels, graded compression ultrasonography was performed from sub-xiphoid to the umbilical zone. The superior mesenteric artery (SMA), which originates from the anterior wall of the aorta, was identified along with its echogenic walls. The superior mesenteric vein (SMV) was identified by following the portal vein confluence caudally until the direction of the mesenteric vein was determined.

A normal superior mesenteric vein (SMV) is located on the right side of the artery. They continue caudally in the middle of the abdomen, parallel to the aorta. In inversion mesenteric vessels, the superior mesenteric vein (SMV) is located on the left side of the artery. In deviation mesenteric vessels, the mesenteric vessels pathway is abnormal and moves to the right or left side of the abdomen. Then, sonographic findings of malrotation were evaluated, including inversion of mesenteric vessels and deviation (abnormal pathway) of mesenteric vessels. Finally, the definitive diagnosis was made according to the gold standard method, which was surgery. Moreover, all the patients received upper GI contrast study and the results were also compared with the results of surgery.

2-4. Data Analyses

The data were analyzed by IBM SPSS software version 22.0 (Chicago, IL, USA). Descriptive statistics were used to describe the findings. To compare the diagnostic performance of sonographic findings in children with and without malrotation, sensitivity, specificity, Positive Predictive Value (PPV) and Negative Predictive Value (NPV) were calculated. These results were also assessed for upper GI series.

3- RESULTS

Totally, 154 children with suspected intestinal malrotation, based on signs and symptoms, were included in this study. The study population was 82 males and 72 females, with a mean age of 22 months ranging from 1 month to 13 years. Among 154 patients, 107 were confirmed to have malrotation; the other final diagnoses are shown in **Table 1**.

Table 2 illustrates the core ultrasonographic findings and upper GI series in patients with and without malrotation. The inversion of the SMA and SMV only detected 63 out of the 107 cases with malrotation, with a sensitivity of 58.9%. The highest sensitivity was observed in deviation of mesenteric vessels, with a sensitivity and specificity of 90% and 17%, respectively. Overall, ultrasonography identified 79 out of the 107 patients with malrotation, with a sensitivity of 74%. On the other hand, the upper GI series had the highest specificity (87.5%) and also a sensitivity (82.5%) higher than that of the ultrasonography.

Out of 87 patients with malrotation, 22 cases (25%) had normal vessels in the proximal part. In other words, in 25% of patients, mesenteric vascular disorder can be detected only in the distal part.

Table-1: Patients' characteristics and final diagnoses (N_{total}= 154)

Variable		N (%)
Gender	Male	82 (53.25)
	Female	72 (46.75)
Diagnosis	Malrotation	107 (69.48)
	Enteric atresia or obstruction	13 (8.44)
	Diaphragmatic hernia	9 (5.84)
	Space-occupying lesions	10 (6.49)
	Enteric adhesions	5 (3.25)
	Annular pancreas	2 (1.30)
	Paraduodenal hernia	1 (0.65)
	Preduodenal portal vein	1 (0.65)
	False positive	6 (3.90)

Table-2: Diagnostic performance of ultrasonographic signs and upper GI series in children

Variables		Malrotation		Sensitivity	Specificity	PPV	NPV
		Positive	Negative				
Inversion of SMV/SMA	Yes (n=93)	63	30	59%	36%	68%	28%
	No (n=61)	44	17				
deviation of mesenteric vessels	Yes (n=110)	71	39	90%	17 %	64.5%	50%
	No (n=16)	8	8				
Upper GI series	Yes (n=33)	32	1	82.5%	87.5%	97%	50%
	No (n=14)	7	7				
Ultrasonography	Yes (n=126)	79	47	74%	-	63%	-
	No (n=28)	28	0				

SMA: superior mesenteric artery; SMV: superior mesenteric vein; GI: gastrointestinal; PPV: positive predictive value; and NPV: negative predictive value.

4- DISCUSSION

Malrotation is one of the common congenital disorders that mainly affects neonates; however, it may have a presentation in adulthood, too (15). As malrotation can result in intestinal necrosis, gangrene, peritonitis, and other life-threatening complications such as life-time dependency on parenteral nutrition, cases should be diagnosed as soon as possible (6, 7, 16). Thus, studies have focused on finding a suitable and accessible method for diagnosing malrotation.

Traditionally, contrast media studies were popular in the case of diagnosing malrotation. The upper GI study of patients with malrotation is characterized by duodeno-jejunal flexure at the right of the midline, the presence of proximal jejunal loops in the right hemi-abdomen, and the whirlpool sign. However, these are not always easy to find, and sometimes the physician cannot rely on these findings (9, 16). Moreover, the whirlpool sign is usually present in patients with volvulus and is rarely found in mere malrotation (17). Furthermore, as the cecum is mobile, the position of this organ cannot be a reliable marker for malrotation. Even in around 20% of the malrotation cases, the position of the cecum is normal and then, barium enema cannot be suitable for the diagnosis of malrotation (18). Moreover, contrast medium studies pose radiation and harm to patients (8, 9).

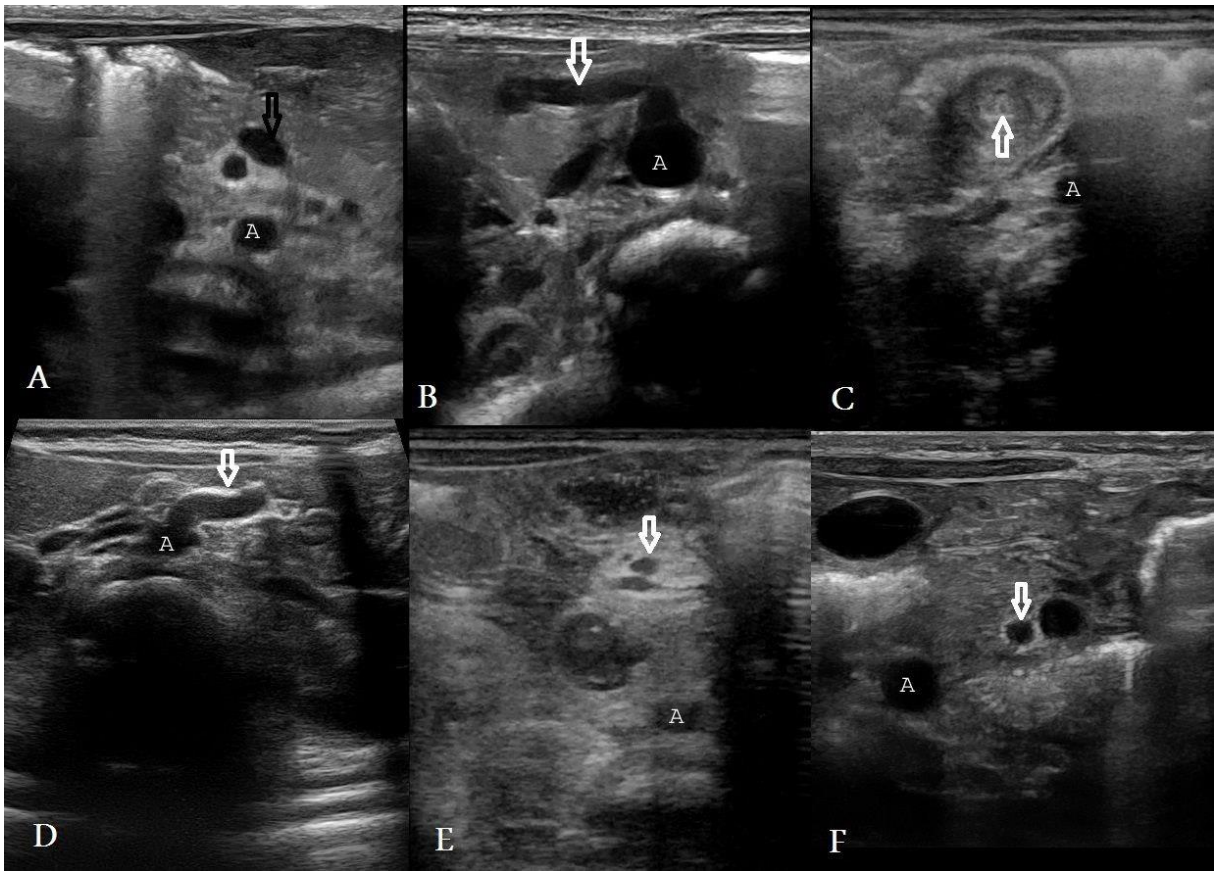

Fig. 1: The types of mesenteric vessels abnormalities in malrotation patients: (A) Inversion of mesenteric vessels alone. B) Deviation (abnormal pathway) of mesenteric vessels to the right side alone. C) The whirlpool sign. D) Left side deviation of mesenteric vessels in the proximal part. E) Right side deviation of mesenteric vessels in distal part. F) Inversion and deviation of mesenteric vessels to the left side in distal part. SMA is marked with white arrows and SMV is marked with black arrows. A: aorta

Our study reported a sensitivity and specificity of 82.5% and 87.5% for upper GI study, respectively. Moreover, the PPV of this modality was 97% and its NPV was 50%. These results were closely similar to another previously reported study by Taghavi et al (19). They reported a sensitivity of 82.5%, specificity of 85.7%, PPV of 97%, and NPV of 46%. In another study by Zhang et al. (13), sensitivity of 92%, specificity of 100%, NPV of 93%, and PPV of 100 were reported for an upper GI study. Of course, the age range of this study was 1 to 31 days and the age range of our study was 1 month to 13 years; thus, the differences in the results should be due to these age differences.

Today, it seems that sonography can take the place of the upper GI series in diagnosing malrotation. In fact, this modality has many advantages, including the absence of harmful radiation and bedside assessment of the patients (20-22). There are different sonography findings that are representative of malrotation. We assessed the diagnostic value of two of these findings, including inversion of SMV/SMA and deviation (abnormal pathway) of mesenteric vessels. In normal anatomy, the SMV is positioned on the right side of the SMA; however, the condition is vice versa in a patient with malrotation as inversion of SMV/SMA (16). In our study, this finding showed a

sensitivity of 59%, specificity of 36%, PPV of 68%, and NPV of 28%. However, Zhou et al. (14), reported higher values for these findings. Accordingly, the calculated sensitivity, specificity, PPV, and NPV were 100%, 97.6%, 95.8%, and 100%, respectively.

The diagnostic value of deviation (abnormal pathway) of mesenteric vessels was also assessed in our study. Deviation of mesenteric artery can be found as a right side or left side turn of the mesenteric artery. We reported a sensitivity of 90%, specificity of 17%, PPV of 72%, and NPV of 50%. However, this finding is not exclusively indicative of malrotation and can be a sign of different disorders like diaphragmatic hernia, masses, vessels anomalies, adhesions, and obstructions (23). The low specificity can be addressed in this regard.

In fact, a summation of different sonography findings seems to have a good diagnostic value. Zhou et al. (14), also combined 3 sonography findings, including inversion of SMV/SMA, the whirlpool sign, and an intraperitoneal transverse duodenum. They reported sensitivity, specificity, PPV, and NPV of 100%, 97.9%, 95.8%, and 100%, respectively. However, we used deviation of mesenteric artery instead of intraperitoneal transverse duodenum, to which the differences between our results and theirs can be partly related. Moreover, whirlpool sign is mainly indicative of volvulus, which is a result of malrotation and the combination of this sign with other signs in detection of malrotation yields a higher false sensitivity.

Overall, the reposted sensitivity and positive predictive value of sonography in our study were 74% and 63%, respectively. Taghavi et al. also reported a sensitivity and specificity of 82% and 54.5% for sonography, respectively (19). They focused on the findings related to the mesenteric artery, including inversion of mesenteric vessels, deviation of mesenteric vessels, duplication of mesenteric vessels, and the anterior position of the mesenteric vein relative to the artery. Zhang et al. reported the sensitivity and PPV to be 100% for this modality (13). However, the method of data collection in their study was different. They analyzed data of all patients diagnosed with ultrasound as malrotation and then confirmed with surgery, while our study included all patients with mesenteric vascular abnormalities on ultrasound and all patients with confirmed malrotation on surgery associated with complete and reliable ultrasound information. Because we retrospectively enrolled sonographic data of all cases with surgically confirmed malrotation too, the reported sensitivity is less than that of the previous studies. Moreover, we assessed patients at a wide range of ages, and this further made our study different from the literature. Due to the type of study, the specificity and NPV of malrotation could not be calculated.

In fact, the available literature tries to propose sonography as a valuable diagnostic method for malrotation. However, more sonography findings should be investigated to improve the diagnostic value of this modality. Contrary to the popular belief, the results of this study show that the deviation of mesenteric vessels is more sensitive to mesenteric vessels inversion for the diagnosis of malrotation (90% versus 59%), although its specificity is low. Moreover, mesenteric artery abnormalities are not accurate enough for malrotation diagnosis. The experienced radiologist with several previous research papers on the same subject and large sample size can be considered as the main strength of our study. However, it is advised that the researchers in this field work on other sonography findings of malrotation patients to make this modality more valuable.

5- CONCLUSION

In conclusion, abnormalities of mesenteric vessels on ultrasound such as inversion and deviation of mesenteric vessels sign are the findings of malrotation. The deviation of mesenteric vessels and the combination of them are the most sensitive findings. The inversion of SMV/SMA seems to be the most valuable sonography finding in our study. Although deviation (abnormal pathway) of mesenteric vessels showed a higher sensitivity compared to the SMV/SMA inversion in detecting malrotation, it is not specific enough for malrotation and can be found in many anomalies. In fact, the proposal and combination of different sonography findings may further help the evolution of sonography in detecting malrotation. Unfortunately, these ultrasound abnormalities aren't seen in all patients with malrotation. Further studies are needed to complete these results.

6- ETHICAL CONSIDERATIONS

The parents or legal guardians completed an informed consent form, according to the World Medical Association Declaration of Helsinki, revised in 2000, Edinburgh. Moreover, the study protocol was fully approved by the Ethics Committee of Mashhad University of Medical Sciences (IR.MUMS.MEDICAL.REC.1398.328).

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