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The Diagnostic Value of Ultrasonography in Pediatric Intestinal Malrotation

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Abstract: ***Background & Objectives: Intestinal malrotation is a congenital disorder, which is characterized by defects in the natural rotational process of intestinal loops. This anomaly is considered important due to its acute and chronic presentations. This study was designed to investigate the diagnostic value of ultrasonographic findings in patients with suspected intestinal malrotation. Materials & Methods: In this prospective, cross-sectional study, performed at Dr. Sheikh Pediatric Hospital of Mashhad during 2009-2014, sixty patients (< 15 years of age) were selected via purposive sampling. The subjects were suspected of having intestinal malrotation, based on upper gastrointestinal findings, surgical manifestations or study the diagnostic accuracy of ultrasound in a group of patients with the inclusion criteria of ultrasonographic features of malrotation (i.e., abnormal pathway of mesenteric vessels such as: the inversion of mesenteric vessels, duplication of the superior mesenteric vein, anterior position of mesenteric vein relative to superior mesenteric artery and whirlpool sign). Results: Among 60 patients, 31 (51.7%) were male, with a median age of 2.8±1.3 years. The most common symptoms were vomiting (45%) and chronic abdominal pain (21.6%), respectively. The rate of associated anomalies was 51.6% and volvulus was reported in 6.6% of patients. Overall, ultrasound evaluations had a sensitivity of 82% in comparison with surgical findings. Conclusion: Based on the findings, ultrasonography of mesenteric vessels is a sensitive diagnostic tool for the detection of intestinal rotational disorders in pediatric populations. Therefore, abnormal ultrasonographic findings should be taken into consideration in suspected patients.***

Keywords: Mesenteric Vessels, Intestinal Malrotation, Children, Sonography.

1. Introduction

Intestinal malrotation is a congenital condition, which is characterized by defects in the embryological process of intestinal rotation or interruption in the attachment and fixation of the intestine to the abdominal wall [1]. Considering the acute and chronic presentations of intestinal malrotation, early diagnosis is of great significance. The presentations of this condition may vary from failure to thrive with non-specific pain to acute volvulus and death [1]. The incidence of intestinal malrotation was reported to be 2.8 cases per 10,000 live births in Hawaii during 1986-1999 [2].

Considering the life-threatening complications of intestinal malrotation, e.g., ischemia and gangrene of the small bowel (which is regarded as a surgical emergency), early diagnosis is crucial. Chronic complications of intestinal malrotation include chronic intestinal obstruction, chronic midgut volvulus and chronic internal hernia [3].

Many patients with a confirmed diagnosis of acute volvulus normally experience a period of non-specific symptoms, which is also associated with frequent misdiagnoses. Therefore, both radiologists and clinicians should be familiar with all the presentations of intestinal malrotation.

Clinical diagnosis of intestinal malrotation is quite challenging, particularly in adolescents and adults. This is due to the fact that disease presentations including abdominal pain and vomiting in these age groups are often non-specific and rarely lead to the clinical suspicion of malrotation [4].

Considering the low prevalence of intestinal malrotation, there is a great deal of controversy about the diagnostic modalities and their diagnostic values. The evaluation of intestinal malrotation by upper gastrointestinal series, as a diagnostic modality, has been emphasized in many studies. Since in a significant percentage of children (15%), the position of cecum is variable due to its mobility, barium enema is less practical in the diagnosis of intestinal malrotation, compared to other modalities [5].

In recent years, considering the features of ultrasonography, i.e., lack of ionizing radiation, non-invasiveness and its feasibility at bedside, this modality has been suggested as an appropriate method for diagnosing intestinal malrotation [6]. Early diagnosis of malrotation with or without volvulus is of great importance. On the other hand, ultrasound is a relatively inexpensive and available method without any serious complications. Therefore, we designed this study to evaluate the diagnostic value of sonographic findings in children suspected of intestinal malrotation.

2. Materials and Methods

This descriptive, cross-sectional study was performed at Doctor Sheikh Pediatric Hospital during 2009-2014 after being approved by the Institutional Review Board of Mashhad University of Medical Sciences, Mashhad, Iran.

Overall, 60 patients under 15 years of age were enrolled in this study. The subjects were all suspected of intestinal malrotation, based on the following findings: ultrasonographic findings suggestive of malrotation (i.e., the inversion of mesenteric vessels, abnormal pathway of mesenteric vessels, duplication of mesenteric veins, anterior position of mesenteric vein relative to mesenteric artery and whirlpool sign), suspected barium enema findings (i.e., abnormal position and course of duodenojejunal flexure, abnormal cecal position and corkscrew appearance of the small intestine) or surgical presentations suggestive of intestinal malrotation.

In cases with suspected ultrasonographic features or upper gastrointestinal findings, surgical procedure was performed as necessary. Ultrasonographic examinations were performed within the first two hours of hospital admission by two experienced radiologists with more than five years of experience in the field of pediatric radiology.

Gray-scale ultrasonography was performed, using sonographic devices including MyLab 50 (Esaote, Italy) and Sonoline Adara (Siemens, Germany). Also, 7.5-12 MHz linear and 3.5 MHz curved probes were used for gray-scale ultrasound examinations. The subjects were scanned in the supine position with the transducer positioned in the midline of the anterior upper abdomen.

Moreover, graded compression sonography was performed for detecting the location and direction of mesenteric vessels. The superior mesenteric artery (SMA) was identified along with its echogenic walls, originating from the anterior wall of the aorta. The superior mesenteric vein (SMV) was localized by tracing the portal vein confluence and moving caudally until the direction of mesenteric vein was identified. The vessels were considered to be inverted in case SMV was oriented towards the left side of SMA (left-sided SMV).

Upper gastrointestinal (UGI) series was performed with barium, which was consumed orally or via a nasogastric tube. The positions of the stomach, duodenum, duodenojejunal junction, jejunum and cecum were assessed. Moreover, contrast study findings related to intestinal malrotation such as the location of ligamentum teres (the position and course of duodenojejunal flexure), the location of cecum and corkscrew appearance of the small intestine were analyzed.

For patients with ultrasonographic or clinical findings suggestive of intestinal malrotation, surgery was performed when indicated; overall, 60 patients underwent surgical procedures. Finally, the ultrasound findings were compared with the final diagnosis of patients.

The obtained data were analyzed by SPSS version 16 (Chicago, IL, USA). P-value less than 0.05 were considered statistically significant.

3. Results

In total, 60 patients with suspected intestinal malrotation, based on ultrasonographic features, UGI series or surgical findings, were enrolled in this study. Overall, 31 (51.7%) and 29 (48.3%) subjects were male and female, respectively. The subjects were within the age range of 2 days to 14 years, with the mean age of 2.8±1.3 years. In our study population, the most common symptoms were vomiting (45%) and chronic abdominal pain (21.6%), respectively. The incidence rate of associated anomalies was 50.8% and volvulus was reported in 6.6% of subjects. Patients' clinical findings are summarized in table 1.

Table 1: The clinical symptoms of subjects with intestinal malrotation

Symptoms	Number (%)
Growth retardation	5 (8.3%)
Vomiting	27 (45.0%)
Abdominal pain	13 (21.6%)
Restlessness	5 (8.3%)
Asymptomatic	4 (6.6%)
Respiratory distress	4 (6.6%)
Cloacal anomaly	1 (1.6%)
Subcutaneous mass	1 (1.6%)

According to Chi-square test results, the primary complaints of male and female subjects were not significantly different at a significance level of 5% (P=0.471). The most common symptoms in patients younger than one year of age were restlessness and vomiting (47.8%). Moreover, growth retardation, abdominal pain and vomiting (63%) were the most common symptoms among children, aged 1-5 years. Also, in patients above five years of age, abdominal pain and growth retardation were found to be the most prevalent symptoms (72%). There was a significant difference between different age groups (<1 year, 1-5 years, and >5 years) in terms of presenting symptoms (P=0.01). The ultrasonographic findings and final diagnoses of patients are summarized in table 2.

Table 2: Comparison of ultrasonographic findings and final diagnosis in patients

Ultrasonographic findings	Number (%)	Final diagnosis
Inversion of mesenteric vessels	45 (75.0%)	Intestinal malrotation (n=41) Jejunal atresia (n=1) Choledocal cyst (n=1)
Abnormal pathway of mesenteric vessels	2 (3.3%)	Gastric volvulus (n=1) Duodenal web (n=1)
Duplication of mesenteric vein	2 (3.3%)	Normal
Anterior position of the mesenteric vein relative to the artery	1 (1.6%)	Normal
Normal	10 (16.0%)	Intestinal malrotation (n=9) Annular pancreas (n=1)

The final diagnosis of intestinal rotational disorders was confirmed in 50 cases (83%). According to the results, the sensitivity of sonography was estimated at 82% with a specificity of about 10% and positive predictive value of 84%. It should be noted that the hospital in which we performed this study is a tertiary center where the diagnosis of almost every admitted patient was almost definitive; this might have in fact resulted in the low specificity of sonographic findings.

Intestinal malrotation was reported in none of the cases with duplicated mesenteric vein, abnormal course of

mesenteric vessels or the anterior location of SMV relative to SMA. The abnormal direction and the inverted course of mesenteric vessels were reported in four cases, who suffered from other gastrointestinal anomalies such as duodenal atresia, gastric volvulus, duodenal web and choledochal cyst.

Intestinal malrotation disorders were associated with other congenital anomalies in 51.6% of patients. The most common anomalies were gastrointestinal anomalies (e.g., annular pancreas and small intestine duplication), anomalies associated with the diaphragm (e.g., Bochdalek hernia and sequestration) and neuromuscular disorders (e.g., myelomeningocele, scoliosis and neurogenic bladder). The whirlpool sign was reported only in four patients (6.6%), which was compatible with the surgical diagnosis of midgut volvulus.

4. Discussion

The process of embryological rotation of gastrointestinal system in a normal embryo is complicated. Consequently, various studies have been performed to specify this process, leading to the development of the theory of intestinal rotation around SMV. Overall, a wide range of congenital abnormalities such as gastroschisis, omphalocele and diaphragmatic hernia lead to rotational disorders and intestinal malrotation, which is itself associated with serious complications and high morbidity and mortality rates [7].

In our study, the most common clinical symptoms of intestinal malrotation were vomiting (45%) and chronic abdominal pain (21.6%), respectively. On the other hand, in a study by Nahvi et al., abdominal distention and bilious vomiting were the most common symptoms among the evaluated neonates [8]. In our study, the rate of associated anomalies was estimated at 51.6%, and the most common disorders included gastrointestinal disorders (e.g., annular pancreas and duplication of the small intestine), diaphragmatic anomalies and neuromuscular disorders, respectively. The high prevalence of associated anomalies in our patients, especially rhabdomyosarcoma and leukemia reported in two patients, is a noteworthy issue, which could be related to the sample size of this study.

In the study by Nahvi et al. [8], congenital disorders were reported in 46% of patients and intestinal anomalies such as intestinal atresia, imperforated anus and duodenal web were the most common of all. Also, in a study by Murphy, other congenital anomalies were reported in 14 children (30%) [7]. Overall, gastrointestinal anomalies such as Bochdalek hernia, annular pancreas, jejunal atresia, duodenal web, choledocal cyst and intestinal malrotation are the main causes of inversion in mesenteric vessels.

In the past, UGI barium study was used as a routine and optimal diagnostic method for evaluating intestinal malrotation [17, 18]. In several recent studies [10, 11], this modality has been shown to have a relatively high missing rate in the diagnosis of intestinal malrotation or even volvulus. Therefore, the use of other diagnostic modalities such as ultrasonography and CT scan for a more accurate diagnosis of intestinal malrotation has been highlighted more than UGI series [12, 13].

In normal abdominal ultrasonography, SMV is located at the right side of SMA (Fig. 1A). Ultrasonographic findings in patients with intestinal malrotation indicated the inversion of mesenteric vessels (Fig. 1B), whereas torsion in the vein around the artery (whirlpool sign) could be observed in midgut volvulus (Fig. 1C). According to our findings, these sonographic findings had a sensitivity of 84%, specificity of 10% and positive predictive value of 82% in diagnosing intestinal malrotation.

In a study by Orzech et al. [9], the sensitivity and specificity of sonography in the diagnosis of intestinal malrotation were 86.5% and 74.7%, respectively. These results were inconsistent with the findings reported in the present study since our hospital is the only referral center in Khorasan province in northeast of Iran, where almost all admitted patients are highly suspected of intestinal malrotation.

The diagnostic value of whirlpool sign in detecting midgut volvulus was significant in our study, which was consistent with the findings reported by Chao et al. and Pracros et al. [3, 14]. The high sensitivity of sonography in the diagnosis of intestinal malrotation and even midgut volvulus in our research and other similar studies indicates that almost all cases of malrotation can be easily diagnosed via sonography; moreover, the failure rate of this technique is often insignificant.

Although the sensitivity of ultrasound in detecting malrotation was very high in our study and similar research (13), the majority of mistakenly reported cases in our study had common variations in mesenteric vessels, based on repeated sonography. The variations of mesenteric vessels in our study were as follows: the anterior location of the cranial part of SMV relative to SMA, the anterior position of the cranial part of SMV relative to the left side of SMA (Fig. 2A) and duplication of SMV in the cranial part (Fig. 2B), especially if the right vein was smaller than the left one (Fig. 2C). To avoid such errors, caudal tracing of our scanning revealed normally located vessels. It should be mentioned that the variations of mesenteric vessels have been extensively reviewed in previous studies [15, 16].

Another interesting point in our study was the effect of the abnormal direction of mesenteric vessels on the diagnosis of other gastrointestinal disorders such as internal hernia or Bochdalek hernia. In a patient in whom the direction of mesenteric vessels was towards the left anterior side of the abdomen, left-sided Bochdalek hernia was detected by UGI contrast study (Fig. 3). In another patient with the unusual direction of mesenteric vessels towards the right side of the abdomen, internal hernia was reported during surgery. These cases reveal the fact that the position and direction of mesenteric vessels should be considered during pediatric ultrasonography.

Furthermore, in our study, the diagnostic value of whirlpool sign for midgut volvulus was significant, since all patients with ultrasonographic whirlpool sign were diagnosed with midgut volvulus in surgery. This was consistent with the findings reported by Chao et al. and Pracros et al. [3, 14]. As a result, sonographic whirlpool

sign had high sensitivity and specificity in the diagnosis of midgut volvulus.

In general, sonography is a safe and widely available diagnostic method, with high sensitivity for detecting life-threatening abdominal anomalies such as intestinal malrotation, volvulus or even obstruction and hernia in the upper abdomen. Meticulous evaluation of the location and direction of mesenteric vessels, normal variations in the pathway of mesenteric vessels and sonographic features of malrotation and volvulus can easily reveal these abnormalities. The absence of ionizing radiation in sonography and its feasibility at patient's bedside are some of the important advantages of this modality.

We performed this study at a tertiary hospital, where children with advanced symptoms were admitted and their diagnoses were nearly definitive; this was in fact one of the limitations of the present study. Another limitation of our research was the high incidence of abdominal anomalies in subjects, which was unexpected and could affect the quality of ultrasound images and clinical findings. Overall, our study population was of a modest size. Therefore, larger prospective studies, along with a meta-analysis of the findings of different studies, could be very helpful.

5. Conclusion

Sonography of mesenteric vessels is a sensitive diagnostic method for detecting intestinal rotational disorders in children. Also, abnormal ultrasonographic findings should be evaluated in suspected patients.

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Figures:

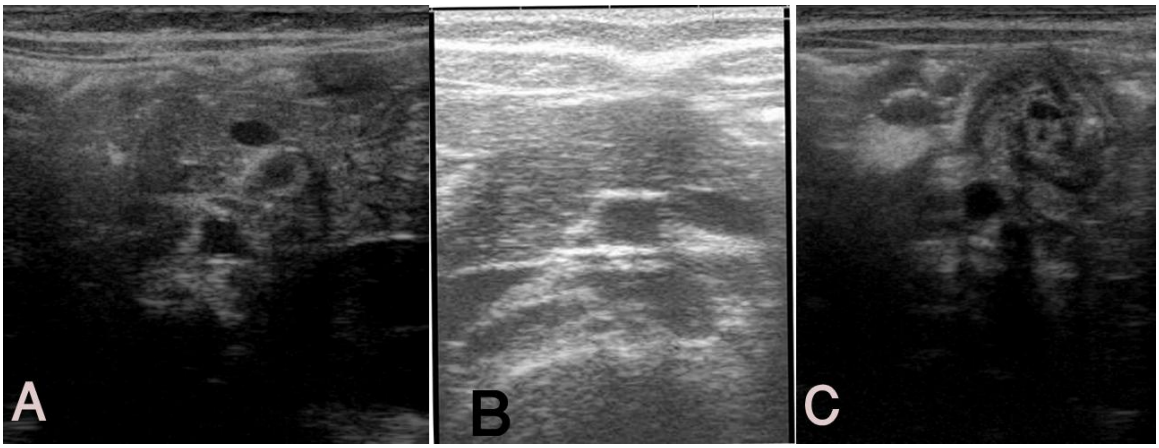

Figure 1: A) The normal orientation of SMA and SMV in a normal population; B) the inversion of SMA and SMV in patients with malrotation; C) whirlpool sign in midgut volvulus

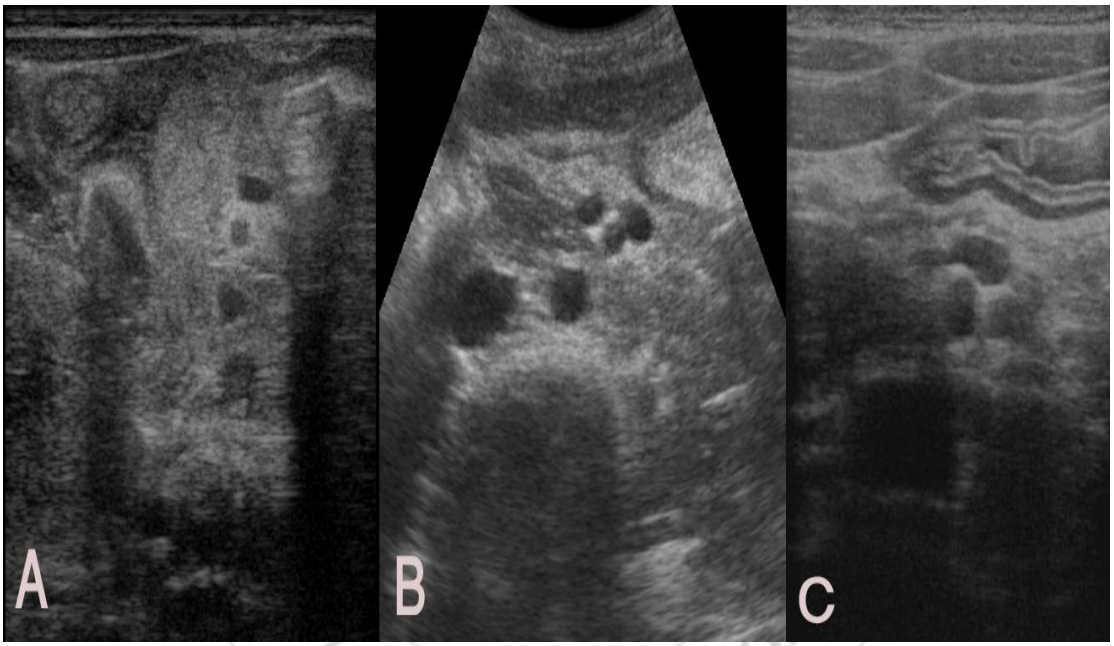

Figure 2: Variations of mesenteric vessels: A) left anterolateral location of the cranial part of SMV relative to SMA; B) duplication of SMV in the cranial part; C) duplication of SMV with the right vein smaller than the left one

Figure 3: The abnormal pathway of mesenteric vessels in Bochdalek hernia