Indian J Gastroenterol (January–February 2017) 36(1):27–31
DOI 10.1007/s12664-017-0729-5

ORIGINAL ARTICLE

Color Doppler—An effective tool for diagnosing midgut volvulus with malrotation

Bindey Kumar[1] · Manish Kumar[2] · Prem Kumar[3] · Amit Kumar Sinha[1] · Utpal Anand[4] · Anil Kumar[5]

Received: 9 October 2016 / Accepted: 5 January 2017 / Published online: 26 January 2017

Abstract
Background Malrotation with midgut volvulus is a common surgical emergency in children. A diagnostic tool for malrotation with characteristics ideal in emergency settings such as non-invasiveness and rapidity remains a keenly debated issue among surgeons and radiologists alike.
Methods Fifty-two pediatric patients of suspected malrotation with midgut volvulus were studied between 1998 and 2016. All patients underwent plain abdominal X-ray and Color Doppler; contrast upper gastrointestinal (GI) studywas done in some patients. All cases were operated and diagnosis was confirmed. A subset of 60 pediatric patients with nonspecific GI complaints were also examined to see relative position of superior mesenteric vein (SMV)/superior mesenteric artery (SMA) in control population.
Result A total of 52 suspected cases of malrotation were admitted from May 1998 to November 2015, 43 had inversion of SMA/SMV and nine had SMV anterior of SMA in Color Doppler. All 43 cases of inversion of SMA/SMV were cases of malrotation after surgical confirmation; while five out of nine cases of SMV anterior to SMA had malrotation.
Conclusion In appropriate clinical settings, Color Doppler documenting the reversal or aberrant SMV/SMA axis is not only predictive but also diagnostic of malrotation of gut.

Keywords Mesenteric vessels · Ultrasonography · Upper GI series

Introduction

Malrotation represents an interrupted embryological process. In the fourth to fifth week of fetal development, bowel extrudes into umbilical cord associated with 90^0 anti-clockwise rotation and as it returns back to the peritoneal cavity by eighth to tenth week, it undergoes further counter-clockwise rotation of 180^0, thus completing a total of 270^0 counter-clock rotation bringing the dudenojejunal flexure posterior and to the left of superior mesenteric artery (SMA), while cecum lies in right lower quadrant. However, incomplete intestinal rotation results in dudenojejunal flexure lying to the right of midline, cecum on left, a narrow mesentery devoid of fixation, and peritoneal bands passing from the cecum to the right side across the duodenum. This pathological entity is predisposed to torsion and obstruction as the narrow mesentery may twist resulting in midgut volvulus [1]. Other forms of rotational anomalies include non-rotation where duodenum passes inferiorly on right without rotation, and reverse rotation where duodenum passes in front of SMA.

Midgut volvulus presents as a surgical emergency whose diagnosis if not established early, leads to catastrophic consequences [2]. Traditionally, upper gastrointestinal (GI) series is considered the diagnostic modality of choice in malrotation [3]; however, the complexities of this embryological enigma

✉ Manish Kumar
singh.manish.15@gmail.com

[1] Department of Pediatric Surgery, All India Institute of Medical Sciences, Phulwari Sharif, Patna 801 507, India

[2] Department of Pediatrics, All India Institute of Medical Sciences, Phulwari Sharif, Patna 801 507, India

[3] Department of Radiology, All India Institute of Medical Sciences, Phulwari Sharif, Patna 801 507, India

[4] Department of Gastrosurgery, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna 800 014, India

[5] Department of Surgery, All India Institute of Medical Sciences, Phulwari Sharif, Patna 801 507, India

has egged surgeons and radiologists alike to look for a diagnostic tool with requisite sensitivity and specificity along with characteristics ideal in emergency settings such as non-invasiveness and rapidity [4]. Color Doppler study with its portability and accessibility may fit the description of an optimum modality of diagnosis in midgut volvulus with malrotation. The signs suggestive of malrotation used in Color Doppler are inversion superior mesenteric vessels and whirlpool sign (WS) which refer to the sonographic appearance of the superior mesenteric vein (SMV), along with the bowel and mesentery, wrapping in a clockwise fashion around the SMA in cases of volvulus.

This study aims to examine the efficacy of Color Doppler signs like relative positions of SMV and SMA and WS in diagnosing midgut volvulus with malrotation.

Methods

Fifty-two pediatric patients of suspected malrotation with midgut volvulus were studied between May 1998 and November 2015. All cases had common clinical presentation of persistent vomiting where vomitus initially had gastric content followed by bilious character. Plain X-ray abdomen in erect posture showed paucity of gases. All patients underwent Color Doppler as per the pre-defined protocol while contrast upper GI study was done in selected cases.

All cases were operated and diagnosis was confirmed. Rotational anomalies were defined based on abnormalities documented during surgery.

- Malrotation—Duodenojejunal flexure on right and cecum lying to the left of midline
- Nonrotation—Duodenum on right and colon left of midline
- Reverse rotation—Duodenum in front of SMA and colon lying anterior to mesentery

A subset of 60 pediatric patients with nonspecific GI complaints also underwent Color Doppler to see relative position of SMV/SMA in control population.

The pre-defined protocol of Color Doppler ultrasonography (USG) required no patient preparation. USG was done on GE Logiq E9 Color Doppler 3D ultrasound in Department of Radiodiagnosis. High resolution linear probe was used for infant and toddlers while in older children curvilinear probe was used. USG was done with patient in supine position. The basic radiological principles involved in this technique included identification of origin of SMA from abdominal aorta and viewing in the short axis in transverse section the orientation of mesenteric vessels in retroperitoneum. Normally, SMV is seen on the right side of SMA. Rotational anomalies bring about aberration in SMV/SMA relationship in the form of SMV situated anterior or ventral to SMA or at its left side. The characteristic imaging feature of midgut volvulus is "whirlpool sign" in which SMV is seen encircling SMA. This whirling of mesenteric vessels happens in a clockwise manner and therefore represents inversion of SMA/SMV relationship. Loss of normal orientation of SMA–SMV axis on Color Doppler imaging may also be suggestive of midgut volvulus.

Subjects with inversion of SMA/SMV i.e. SMV left of SMA along with those where SMV was seen anterior to SMA on Color Doppler were noted. Subsequently, data was analyzed to assess whether the aberration of SMA/SMV orientation detected by Color Doppler was predictive of malrotation with midgut volvulus.

Results

A total of 52 suspected cases of malrotation were admitted from May 1998 to November 2015. The median age of children was 12 months (IQR 1.6 to 39 months). There were 42 males and 10 females with a male to female ratio of 4.2:1. Color Doppler was performed on all suspected cases for diagnosis of malrotation which were later confirmed by surgical procedure. Out of 52 suspected cases, 43 had inversion of SMA/SMV on Color Doppler, and in nine cases, SMV was seen anterior to SMA on Color Doppler. Out of 43 cases of inversion of SMA/SMV, all 43 were cases of malrotation with midgut volvulus after surgical confirmation. Out of nine cases where SMV was anterior of SMA, five were diagnosed as malrotation after surgical confirmation. A group of 60 control subjects, who presented with some GI complains to the clinic, were also subjected to diagnosis with sonography. None of them were found to have any core signs like inversion of SMA/SMV or anterior of SMA. The median age of the control subjects was 8 years (IQR 1.25 to 16 years), and male to female ratio was 3:2 with 36 males and 24 females. Result has been summarized in Table 1 and Table 2.

Table 1 presents 43 cases of inversion of SMA/SMV compared to surgical confirmation which is considered as "Gold Standard" for confirmation of malrotation. The sensitivity and specificity of sonographic test was 100%, respectively, i.e. all the 43 cases with SMV/SMA were diagnosed as cases of malrotation with midgut volvulus by surgical confirmation while inversion of mesenteric was not seen in any of the controls. The positive predictive value (PPV) of sonographic test was 100% i.e. all who had signs of SMV/SMA by sonographic test were diagnosed for malrotation. Hence, the sonographic test had 100% predictive ability for diagnosis of malrotation. The negative predictive value (NPV) of the test was also 100% indicating no disease among the control subjects. The accuracy of the sonographic test was 100% indicating a high validity.

Table 1 Comparison of Color Doppler test results (SMA/SMV inversion) with surgical procedure

Inversion of SMV/SMA by sonography	Malrotation confirmation by surgical procedure			
	Yes	No	Total	
Yes	43	0	43	PPV = 100%
No	0	60	60	NPV = 100%
Total	43	60	103	Accuracy = 100%
	Sensitivity = 100%	Specificity = 100%		

SMV superior mesenteric vein, *SMA* superior mesenteric artery, *PPV* positive predictive value, *NPV* negative predictive value

Table 2 presents nine cases where SMV was seen anterior of SMA compared to surgical confirmation. Out of nine cases of anterior of SMA, five were diagnosed as malrotation with midgut volvulus after surgical confirmation. The sensitivity of sonographic test was 100% i.e. all the five cases were diagnosed as cases of malrotation by surgical confirmation whereas aberration of mesenteric vessels with SMV anterior to SMA was not present in any control subject. The PPV of sonographic test was 55.5% i.e. the cases who had signs of anterior of SMA by sonographic test, only 55.5% of them were correctly diagnosed for malrotation. Hence, the sonographic test had 55.5% predictive ability for diagnosis of malrotation in cases of anterior of SMA. The NPV of the test was 100% indicating no disease among the control subjects. However, the accuracy of the sonographic test was 94.2% indicating a high validity. Proportion of malrotation was significantly higher among the cases presented with anterior of SMA as compared to control subjects ($p < 0.001$).

Discussion

From Václav Treitz's description "On a new muscle in the human duodenum, over elastic sinews and some other anatomical relations" [5] to William E Ladd's procedure of "restoring an earlier state of embryologic development" [6]; malrotation of gut has always been an enigmatic entity for surgeons and radiologists.

Imaging in midgut volvulus with malrotation has evolved from plain X-ray abdomen which may document the obstruction but in many cases may be non-contributory. Later Ladd and his successor Gross stated their preference for contrast enema for diagnosing malrotation [6].

In 1960s, upper GI series emerged as gold standard diagnostic modality in cases of malrotation based on abnormal position of duodenojejunal junction [7]. In early 1980s for the first time, radiologists documented reversal of normal position of SMA and SMV which provided impetus to evaluation of ultrasound as a diagnostic modality for cases of malrotation with midgut volvulus [8]. The signs used in Color Doppler ultrasound are inversion (Fig. 1) or aberration (Fig. 2) of superior mesenteric vessels and whirlpool sign (Fig. 3) which refers to the sonographic appearance of the SMV, wrapping in a clockwise fashion around the SMA, along with the bowel and mesentery, in cases of volvulus. The advantages of rapidity, easy access, portability, and lack of radiations in case of US Color Doppler make it an attractive option for diagnosing midgut volvulus with malrotation where evaluation time is at a premium! [9].

In our study, all cases in which inversion of mesenteric vessels was documented, turned out to be cases of malrotation with midgut volvulus. This finding is in concurrence with multiple other studies who similarly reported the 100% specificity of the finding [10–12]. Majority of cases where SMV

Table 2 Comparison of Color Doppler test results (SMV anterior of SMA) with surgical procedure

SMV anterior of SMA by sonography	Malrotation confirmation by surgical procedure			
	Yes	No	Total	
Yes	5	4	9	PPV = 55.5%
No	0	60	60	NPV = 100%
Total	5	64	69	Accuracy = 94.2%
	Sensitivity = 100%	Specificity = 93.75%		

SMV superior mesenteric vein, *SMA* superior mesenteric artery, *PPV* positive predictive value, *NPV* negative predictive value

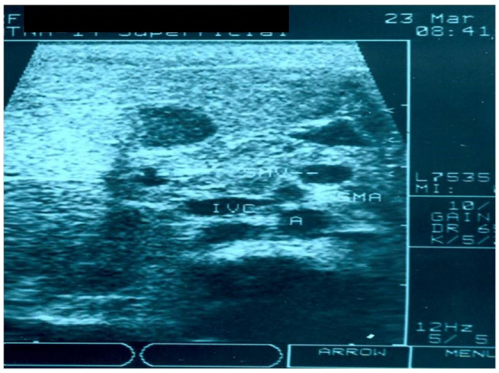

Fig. 1 Color Doppler showing SMA-SMV inversion

was anterior to SMA, our study also had malrotation thus making the sonographic finding of inversion/aberrant mesenteric vessels a reliable predictor of malrotation in appropriate clinical settings.

Few studies have questioned the reliability of ultrasound by citing that similar sonographic finding of mesenteric vessel inversion can be seen in patients with normal midgut rotation, abdominal masses, and distal ileocolic intussusception [13, 14]. However, the control arm of our study, did not document any mesenteric vessel aberrancy on Color Doppler among subjects presenting with non-specific GI symptoms. Thus, the questions regarding reliability are not as robust as suggested by some series. In fact, the upper GI series also suffers from similar ambiguity, as in this modality, the diagnosis of malrotation is based on position of duodenojejunal flexure and therefore, situations such as stomach or small bowel distension, placement of enteric tube may lead to difficulty in interpretation and ambiguity due to distortion of anatomy [15]. Additionally, the procedure puts the child at risk of radiation hazards, risk of hypothermia, and difficult handling. Thus, none of the available radiological modalities are devoid of diagnostic uncertainty, and in this light, the Color Doppler study scores over UGI series specially in terms of feasibility.

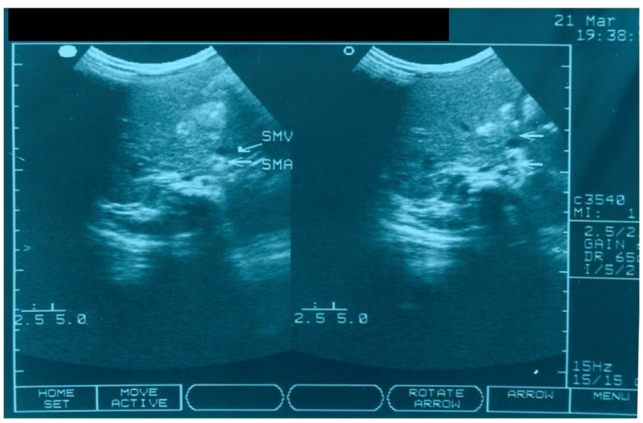

Fig. 2 Color Doppler showing SMV anterior to SMA

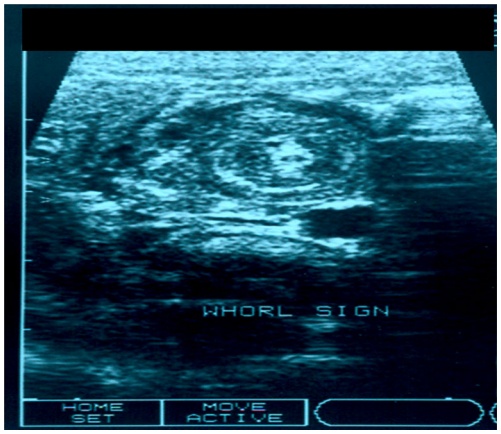

Fig. 3 Color Doppler showing whirlpool sign

The study suffers from the limitation that all cases in this series are classical cases of midgut malrotation, and utility of Color Doppler in cases of non-rotation or reversed rotation could not be examined. Further, the authors recognize that a concurrent comparison with other modalities in all cases such as UGI series and CT abdomen would have given more credence to the claim of Color Doppler being the optimum modality in cases of malrotation with volvulus. Nevertheless, in the appropriate clinical settings, Color Doppler documenting the reversal or aberrant SMV-SMA axis is useful in the diagnosis of midgut volvulus with malrotation.

Acknowledgement Authors acknowledge the contribution of Dr. Alok Ranjan, Asst Prof, Dept of CFM, AIIMS, Patna in the statistical analysis of data.

Compliance with ethical standards

Conflict of interest BK, MK, PK, AKS, UA, and AK declare that they have no conflict of interest.

Ethics statement The authors declare that the study was performed in a manner to conform to the Helsinki declaration of 1975, as revised in 2000 and 2008 concerning human and animal rights, and the authors followed the policy concerning informed consent as shown on Springer.com.

Source of funding None.

References

1. Warner BW. Malrotation. In: Oldham KT, Colombani PM, Foglia RP, eds. Surgery of Infants and Children: Scientific Principles and Practice. Philadelphia: Lippincott-Raven; 1997. p. 1229–40.
2. Lampl B, Levin TL, Berdon WE, Cowles RA. Malrotation and migut volvulus: a historical review and current controversies in diagnosis and management. Pediatr Radiol. 2009;39:359–66.
3. Strouse PJ. Disorders of intestinal rotation and fixation (malrotation). Pediatr Radiol. 2004;34:837–51.

4. Long FR, Kramer SS, Markowitz RI, Taylor GE. Radiographic patterns of intestinal malrotation in children. Radiographics. 1996;16:547–56. discussion 556-60
5. Alford WC Jr. Wenzel Treitz: the man and his "ligament". Surgery. 1963;53:556–62.
6. Ladd WE. Surgical disease of the alimentary tract in infants. N Engl J Med. 1936;215:705–8.
7. Sizemore AW, Rabbani KZ, Ladd A, Applegate KE. Diagnostic performance of the upper gastrointestinal series in the evaluation of children with clinically suspected malrotation. Pediatr Radiol. 2008;38:518–28.
8. Weinberger E, Winters WD, Liddell RM, Rosenbaum DM, Krauter D. Sonographic diagnosis of intestinal malrotation in infants: importance of the relative positions of the superior mesenteric vein and artery. AJR Am J Roentgenol. 1992;159:825–8.
9. Orzech N, Navarro OM, Langer JC. Is ultrasonography a good screening test for intestinal malrotation? J Pediatr Surg. 2006;41:1005–9.
10. Dufour D, Delaet MH, Dassonville M, Cadranel S, Perlmutter N. Midgut malrotation, the reliability of sonographic diagnosis. Pediatr Radiol. 1992;22:21–3.
11. Zerin JM, DiPietro MA. Superior mesenteric vascular anatomy at US in patients with surgically proved malrotation of the midgut. Radiology. 1992;183:693–4.
12. Chao HC, Kong MS, Chen JY, Lin SJ, Lin JN. Sonographic features related to volvulus in neonatal intestinal malrotation. J Ultrasound Med. 2000;19:371–6.
13. Ashley LM, Allen S, Teele RL. A normal sonogram does not exclude malrotation. Pediatr Radiol. 2001;31:354–6.
14. Papadopoulou F, Efremidis SC, Raptopoulou A, Tryfonas GI, Tsikopoulos G. Distal ileocolic intussusception: another cause of inversion of superior mesenteric vessels in infants. AJR Am J Roentgenol. 1996;167:1243–6.
15. Applegate KE, Anderson JM, Klatte E. Intestinal malrotation in children: a problem-solving approach to the upper gastrointestinal series. Radiographics. 2006;26:1485–500.