

Evaluation of the predictability of clinical and radiological findings in the diagnosis of malrotation

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ABSTRACT

Background. To evaluate the predictability of clinical and radiological findings in the diagnosis of malrotation.

Methods. Between 2010 and 2020, children with presumptive diagnosis of malrotation were included. The demographic features, clinical and radiological findings, operative findings and outcome were recorded. The upper gastrointestinal series (UGIS) were evaluated by two radiologists. All parameters were correlated with surgical findings to evaluate the predictability.

Results. Seventy patients were included. The presenting symptom was bilious vomiting in 29 cases (41.4%), and atypical symptoms (non-bilious vomiting, food refusal, etc.) in 40 cases (57%). One of the cases (1.6%) was asymptomatic and diagnosed incidentally during UGIS. 52 cases had abdominal X-ray and 14 (26.9%) of them were normal. Doppler ultrasonography (US) (n=20) revealed evidence of malrotation in 13 cases (65%). The location of duodenojejunal junction (DJJ) in UGIS was compatible with malrotation in 33 cases. 48 (61%) cases underwent surgical exploration; 35 cases had malrotation and seven cases had midgut volvulus. Median follow-up time was one year (0.5-7 years). Volvulus has recurred in one case and another case operated for volvulus died because of short bowel syndrome. The statistical analysis for predictability revealed that bilious vomiting (sensitivity: 57.1%, specificity: 82.1%), Doppler US (sensitivity: 92.3%, specificity: 75%) and right-sided DJJ in UGIS (sensitivity: 96.8%, specificity: 75%) have highest predictability.

Conclusions. The bilious vomiting, Doppler US findings and right-sided DJJ have the highest predictability to confirm the diagnosis. However, presenting with atypical symptoms and having atypical or normal findings in UGIS do not rule out malrotation.

Key words: malrotation, bilious vomiting, treitz ligament, duodenojejunal junction.

Malrotation represents a spectrum of disorders occurring due to abnormal bowel rotation and fixation during embryogenesis. This abnormal bowel development leads to a narrow mesenteric root lying between medially-located duodenojejunal junction (DJJ) and cecum on left side, and peritoneal bands crossing over

duodenum.¹ The most catastrophic scenario of this pathologic condition is twisting of midgut around the narrow mesenteric root causing bowel ischemia, necrosis and eventually sepsis and death. On the other hand, a small number of cases do not become symptomatic throughout their life.¹ Moreover, an important minority of patients experience late-presenting atypical symptoms such as gastroesophageal reflux, intermittent vomiting, chronic and intermittent abdominal pain, and failure to thrive.¹⁻³

Malrotation is seen in 1 in 500 live births and 75% of the cases become symptomatic during neonatal period and up to 90% of the cases are diagnosed during infancy.⁴ The most common

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The abstract of this study was presented as a poster at the 22nd Congress of European Paediatric Surgeons' Association (EUPSA 2021), Athena, 2021.

Received 2nd November 2021, revised 5th March 2022, accepted 10th March 2022.

symptom of these early-diagnosed cases is sudden onset of bilious vomiting. The diagnosis and treatment are obvious in these patients.^{4,5} However, asymptomatic cases and cases with atypical symptoms require radiological examinations for diagnosis. Eventually, a controversy exists regarding the management of incidentally diagnosed malrotations. Although there is still an ongoing disagreement regarding management of asymptomatic cases, most surgeons prefer to perform prophylactic surgery to prevent the risk of emergency situations including volvulus.⁵ Therefore, the predictability of radiologic investigations that may become symptomatic have paramount importance in deciding prophylactic surgery in asymptomatic cases. The most frequently used radiological sign for the diagnosis of malrotation is the evaluation of DJJ with upper gastrointestinal contrast series (UGIS).^{2,6,7} The evaluation and interpretation of the findings in UGIS has a crucial role in the diagnosis since several clinical scenarios such as splenomegaly, gastric distention, and retroperitoneal tumors may mimic malrotation. Furthermore, the variations of DJJ positioning should be kept in mind to prevent false positive or false negative results.^{7,8} In recent decade, Doppler ultrasonography (US) has becoming the first diagnostic tool choice in the diagnosis of malrotation with an advantage of no radiation risk.^{6,9,10} Although there are well-defined sonographic signs of malrotation in the literature, it was also stated that the normally located superior mesenteric vein does not exclude malrotation.⁶ Therefore, we conducted a study reviewing the historical medical records of our cases to evaluate the predictability of clinical and radiological findings in the diagnosis of malrotation.

Material and Methods

We carried out a case series analysis on all cases that were evaluated for suspicion of malrotation between 2010 and 2020. At first, we performed a keyword search of radiology reports including the keywords 'malrotation, DJJ position, Treitz ligament position'. Then, the extracted reports were re-identified for the patients' characteristics and the patients evaluated in the pediatric surgery department were included. The excluded cases were adult ones, the ones that were not admitted to the department of pediatric surgery, and ones in which images of radiological examinations could not be obtained from the radiology picture archiving and communication system.

After approved by the local ethical committee and the local hospital management, we exported 357 records containing the keywords 'malrotation, DJJ position, Treitz ligament position' via the keyword search of radiology reports. We had 184 records after the repeated examinations were excluded. The cases of these records were reviewed from the hospital patient recording system and 80 cases were found eligible for investigation according to the pre-defined inclusion criteria. The images of the radiological examinations could not be found in the hospital system for 10 cases. Therefore, we included a total of 70 cases in the final review of the study.

The medical records of the included cases were evaluated by using the hospital patient recording system. The demographic features, clinical findings, Doppler US results, operative findings and outcome were recorded from the hospital patient recording system. The radiological examinations were re-evaluated by two blind radiologists from the radiology

picture archiving and communication system. Since only the records of abdominal X-ray and UGIS exist in the database, these images of the included cases could be re-evaluated.

For the measurement of predictability, we defined each parameter of the admission symptoms, abdominal X-ray findings, Doppler US findings and UGIS findings separately. The admission symptoms were evaluated in three groups as asymptomatic, symptomatic (bilious vomiting), and atypical symptoms (non-bilious vomiting, intermittent abdominal pain, food refusal, failure to thrive). Abdominal X-ray findings were evaluated as normal if normal bowel gas distribution is seen and pathologic if gasless abdomen, air-fluid levels or bowel gas collected only on one side are seen. Doppler US findings were evaluated as normal if superior mesenteric artery (SMA) and vein (SMV) positions are normal and pathologic if there is a whirlpool sign or SMV is located anterior or on the left side of SMA. Each finding in UGIS was considered as a different parameter. These findings were defined according to the position of DJJ; (a) malrotation, the duodenum and jejunum remain to the right of the spine, (b) malrotation with a corkscrew duodenum and jejunum, (c) malrotation with low position of the DJJ at midline and inferior of duodenal bulb, (d) partial rotation of the duodenum with the DJJ over the right pedicle, (e) normal location of the DJJ in the left upper quadrant. The location of cecum was considered as mobile cecum or located at left side of the abdomen. Then we correlated all parameters with surgical outcome to evaluate predictability separately.

Statistical analysis was performed by using The Statistical Package for the Social Sciences version 23.0 for Windows (SPSS, Inc., Chicago, IL, USA). The descriptive statistics for quantitative data are given in arithmetic mean, standard deviation, median, and minimum-

maximum values. Qualitative data were summarized using frequency and percentages. Pearson Chi-square test or Fisher's Exact test were used to compare categorical variables. Sensitivity and specificity was obtained for each diagnostic parameter. A p value of less than 0.05 was considered to indicate a statistically significant difference. The study was performed under adherence to the Declaration of Helsinki and by approval of the Local Ethical Committee (GO19/400-2019/10-38).

Results

We included a total of 70 cases into the final review of the study. 60% of cases (n=42) were less than 1 year of age at the time of admission, and 15 of them (21.4%) were newborn at admission. The remaining 40% of cases (n=28) were older than 1 year of age at admission. The presenting symptom was bilious vomiting in 29 cases (41.4%) and atypical symptoms (non-bilious vomiting, food refusal, etc.) in 40 cases (57%). Only one case was asymptomatic and Abdominal US was performed for cystic fibrosis work-up and nephrocalcinosis. The US surprisingly revealed that SMV was located anterior to SMA and the patient was diagnosed as malrotation. The diagnosis was confirmed with UGIS.

Abdominal X-ray was performed in 52 cases and revealed normal in 26.9% (n=14). The pathologic findings in abdominal X-ray were abnormal gas distribution in 35 cases (67.4%), air-fluid level in 2 cases (3.8%) and gasless abdomen sign in 1 case (1.9%).

Doppler US was performed in 20 cases and revealed evidence of malrotation in 13 (65%). Among these 13 cases, the whirlpool sign was present in 4 cases (20%) and SMV was located at left side of SMA in 9 cases (45%).

Table I. The summary regarding the management of cases in comparison with admission symptoms and UGIS findings.

	Management			Total
	Medical follow-up (n=22)	Surgery (Ladd procedure) (n=42)	Surgery (Other pathologies) (n=6)	
Admission symptom				
No symptom	1	0	0	1
Atypical symptom	19	18 (V:1)	3	40
Symptomatic (bilious vomiting)	2	24 (V:6)	3	29
UGIS findings				
a. Malrotation with right sided DJJ	3	22	0	25
b. Malrotation with corkscrew sign	0	8 (V:5)	0	8
c. Malrotation with low position of the DJJ at midline	5	5 (V:1)	0	10
d. Partial rotation of the duodenum with the DJJ over the right pedicle	9	6 (V:1)	2	17
e. Normal location of the DJJ	5	1	4	10

UGIS: Upper gastrointestinal series, DJJ: Duodenojejunal junction

The UGIS examination revealed that the location of DJJ was compatible with malrotation in 33 cases (47%). Among these 33 cases, DJJ was located at the right side of the vertebra in 25 cases and at the right side of the vertebra with corkscrew sign in 8 cases. The UGIS revealed malrotation with low position of the DJJ at midline in 10 cases (14.3%) and revealed partial rotation of the duodenum with the DJJ over the right pedicle in 17 cases (24.3%). The location of cecum could be examined in 28 cases revealing left sided cecum in 3 cases (10.7%) and mobile cecum in 13 cases (46.4%).

Forty-eight (61%) cases underwent surgical exploration and 35 of them had malrotation, 7 of them had midgut volvulus and 6 of them had other surgical pathologies (duodenal web, adhesive band, etc.) (Table I). The median follow-up time was 1 year (0.5-7 years). Volvulus has recurred in one of the surgically managed cases and one case operated for volvulus died because of short bowel syndrome.

The parameters including admission age, admission symptom, and pathological findings in abdominal X-ray, Doppler US and UGIS were correlated with surgical findings to measure the predictability of each parameter. The detailed results of the comparison are given in Table II revealing that statistically significant results are found for the parameters including bilious vomiting, Doppler US findings, and positioning of DJJ at right side of vertebra in UGIS. The low position of DJJ at midline and partial rotation of the duodenum with the DJJ over the right pedicle had a non-significant effect on surgical outcome. The normal position of the DJJ in UGIS (n=10) did not exclude malrotation in one case. The statistical analysis for predictability revealed that bilious vomiting (sensitivity: 57.1%, specificity: 82.1%), Doppler US (sensitivity: 92.3%, specificity: 75%) and positioning of DJJ at right side of vertebra in UGIS (sensitivity: 96.8%, specificity: 75%) had the highest sensitivity and specificity values. The sensitivity and specificity of the parameters are given in Table III.

Table II. The comparison of each parameter with surgical findings.

	Malrotation/Volvulus *		
	Present	Absent	Total
Admission age			
<1 age	27	15	42 (60%)
>1 age	13	15	28 (40%)
			p=0.139
Admission symptom			
None or atypical symptom	18	23	41 (58.5%)
Bilious vomiting	24	5	29 (41.5%)
			p=0.001
Abdominal X-ray finding (n=52)			
Normal	6	8	14 (26.9%)
Pathological	26	12	38 (73.1%)
			p=0.093
Abdominal USG finding (n=20)			
Normal	1	6	7 (35%)
Pathological	12	1	13 (65%)
			p=0.001
UGIS finding (n=70)			
a. Malrotation with right sided DJJ	22	3	25 (35.7%)
b. Malrotation with corkscrew sign	8	0	8 (11.3%)
c. Malrotation with low position of the DJJ at midline	5	5	10 (14.3%)
d. Partial rotation of the duodenum with the DJJ over the right pedicle	6	11	17 (24.4%)
e. Normal location of the DJJ	1	9	10 (14.3%)
			p<0.001

*: **present:** malrotation/volvulus was found in surgical exploration, **absent:** cases that did not surgically managed or did not have malrotation/volvulus in surgical exploration. USG: Ultrasonography, UGIS: Upper gastrointestinal series, DJJ: Duodenojejunal junction

Table III. The summary of statistical analysis showing sensitivity and specificity values for each parameter.

	Sensitivity	Specificity
Bilious vomiting	57.1%	82.1%
Abdominal X-ray	81.3%	40%
Doppler US	92.3%	85.7%
UGIS Parameters		
Malrotation with right sided DJJ with/without corkscrew sign	96.8%	75%
Malrotation with low position of the DJJ at midline	83.3%	64.3%
Partial rotation of the duodenum with the DJJ over the right pedicle	85.7%	45%
UGIS revealing mobile cecum	62.5%	66.%
UGIS revealing cecum located at left side	25%	85.7%

US: Ultrasonography, UGIS: Upper gastrointestinal series, DJJ: Duodenojejunal junction

Discussion

The life-threatening consequences of malrotation make clinicians alert for immediate diagnosis of malrotation especially in neonates complaining of sudden onset bilious vomiting. Previous studies showed that the increase in awareness of malrotation and its consequences have caused an increased number of UGIS examinations but did not change the detection rate of malrotation.¹¹ It was reported that malrotation was found in 9% of UGIS of infants having bilious vomiting.¹¹ This challenge occurred not only because of the possibility of other etiologies of bilious vomiting, but also the possibility of atypical and asymptomatic presentation of malrotation. Therefore, radiological examinations become critical in precise diagnosis and consequently early management of malrotation. On the other hand, as Goldman-Yassen et al. stated, the yield of UGIS should be considered while we are performing in cases with non-specific symptoms in order to decrease radiation exposure.¹² In the present study, malrotation was confirmed via surgical exploration in 82% of cases having bilious vomiting. Whereas, malrotation was confirmed in 44% of cases having atypical symptoms. This significant difference also shows an important point that atypical symptoms do not rule out the malrotation.

Since malrotation is the abnormal positioning of intestines due to an embryological defect in intestinal rotation and fixation, the main target of radiological examinations is to show this abnormal positioning of the bowels. The UGIS is still accepted as the gold standard examination method in diagnosis of malrotation. The important landmark that should be examined via UGIS is mainly the location of DJJ and secondly the location of cecum. The findings that are seen in malrotation are; DJJ located at the right side of the left pedicle of vertebra, DJJ located inferior to duodenal bulb, anteriorly located DJJ on lateral images, gathering of

proximal jejunal loops at the right side of abdomen.⁷ Although location of cecum can also be used as a landmark in diagnosis of malrotation, it should be kept in mind that normal location of cecum does not exclude malrotation.⁷ Although these landmarks are well-defined in the literature, there could be subtle findings or several variations that make the diagnosis challenging.

The sensitivity of UGIS is reported as 93 to 100% in the literature. However, false positive results up to 15% or false negative results up to 6% can be seen.^{2,6,7} False positive results can be seen in several clinical situations including splenomegaly, retroperitoneal tumors, gastric over distention, small bowel obstruction and scoliosis that could mimic malrotation in UGIS images.² In the present study, none of these conditions were present in the patients. Nevertheless, malrotation was not confirmed in 12% of cases having right-sided DJJ in UGIS. Several variations of duodenal anatomy should be kept in mind while examining the UGIS such as wandering duodenum, duodenum inversum and mobile duodenum to prevent false positive results.^{7,8} Although the location of DJJ is the gold standard landmark in diagnosis of malrotation, sensitivity and specificity for each separate location of DJJ has not been reported before. In the present study, we calculated sensitivity and specificity for each separate location of DJJ, and found that the highest scores were seen for right-sided DJJ. The DJJ located at midline or inferior to duodenal bulb has lower sensitivity and specificity values with a statistically non-significant difference.

In recent decades, Doppler US has becoming the first choice diagnostic tool in diagnosis of malrotation since it has less radiation risk.^{6,9,10} The diagnostic findings in Doppler US are twisting of SMV around SMA, namely whirlpool sign, and inversion of SMA and SMV course.^{10,13} Although it is dependent on the experience of radiologist, it can also be performed as a fast

diagnostic method called point-of-care US especially in emergency conditions.¹³ However, normal SMA and SMV course does not exclude malrotation¹⁴ as we found in our study. In the present study, we performed Doppler US in 20 cases with high sensitivity (92.3%) but relatively lower specificity (85.7%). These results are not compatible with the ones in some reports revealing 93.8% sensitivity and 100% specificity for Doppler US.⁹ This difference may be due to the small number of cases having non-specific symptoms in that study⁹ in contrast to our study.

The management of clinically and radiologically suspected malrotation cases is obviously surgical exploration especially in emergency conditions. However, elective surgery or watchful waiting is controversial in these cases since elective surgery is not totally innocent and watchful waiting leaves the patient susceptible to catastrophic consequences such as volvulus and intestinal necrosis. In the present study, the long-term complications including recurrent volvulus and short bowel syndrome and death were seen in urgently operated cases. This observation confirms the reports revealing that emergency operation has higher complication rates.⁵ Until further studies will show which patients will become symptomatic and undergo volvulus, we suggest operating all suspected cases to prevent catastrophic consequences of malrotation.

This study has several limitations because of its retrospective nature and small sample size. Despite its limitations, it has some strong points such as examining each separate parameter for its predictability in diagnosis of malrotation. Nevertheless, further prospective studies are needed to confirm our results.

In conclusion, the bilious vomiting, Doppler US findings consistent with malrotation and right-sided DJJ have the highest predictability to confirm the diagnosis. However, presenting with atypical symptoms and having atypical or normal findings in UGIS do not rule out malrotation.

Ethical approval

The study was performed under adherence to the Declaration of Helsinki and by approval of the Local Ethical Committee (GO19/400-2019/10-38).

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: TS, FCT, MH; data collection: OBT, BRU, HNO; analysis and interpretation of results: UEA, MH, TS, FCT; draft manuscript preparation: OBT, HNO. All authors reviewed the results and approved the final version of the manuscript.

Source of funding

The authors declare the study received no funding.

Conflict of interest

The authors declare that there is no conflict of interest.

REFERENCES

1. Langer JC. Intestinal rotation abnormalities and midgut volvulus. *Surg Clin N Am* 2017; 97: 147-159. <https://doi.org/10.1016/j.suc.2016.08.011>
2. Smitthimedhin A, Suarez A, Webb RL, Otero HJ. Mimics of malrotation on pediatric upper gastrointestinal series: a pictorial review. *Abdom Radiol* 2018; 43: 2246-2254. <https://doi.org/10.1007/s00261-018-1537-9>
3. Ezer SS, Oguzkurt P, Temiz A, et al. Intestinal malrotation needs immediate consideration and investigation. *Pediatr Int* 2016; 58: 1200-1204. <https://doi.org/10.1111/ped.13075>
4. Nagdeve NG, Qureshi AM, Bhingare PD, Shinde SK. Malrotation beyond infancy. *J Pediatr Surg* 2012; 47: 2016-2032. <https://doi.org/10.1016/j.jpedsurg.2012.06.013>

5. Covey SE, Putnam LR, Anderson KT, Tsao K. Prophylactic versus symptomatic Ladd procedure for pediatric malrotation. *J Surg Res* 2016; 205: 327-330. <https://doi.org/10.1016/j.jss.2016.06.097>
6. Carol AG, Kavanagh RG, Leidhin CN, Cullinan NM, Lavelle LP, Malone DE. Comparative effectiveness of imaging modalities for the diagnosis of intestinal obstruction in neonates and infants: a critically appraised topic. *Acad Radiol* 2016; 23: 559-568. <https://doi.org/10.1016/j.acra.2015.12.014>
7. Kumbhar SS, Qi J. Fluoroscopic Diagnosis of Malrotation: Technique, Challenges, and Trouble Shooting. *Curr Probl Diagn Radiol* 2020; 49: 476-488. <https://doi.org/10.1067/j.cpradiol.2019.10.002>
8. Long FR, Kramer SS, Markowitz RI, Taylor GE. Radiographic patterns of intestinal malrotation in children. *Radiographics* 1996; 16: 547-560. <https://doi.org/10.1148/radiographics.16.3.8897623>
9. Karaman İ, Karaman A, Çınar HG, Ertürk A, Erdoğan D, Özgüner İF. Is color Doppler a reliable method for the diagnosis of malrotation?. *J Med Ultrason* (2001) 2018; 45: 59-64. <https://doi.org/10.1007/s10396-017-0794-5>
10. Kumar B, Kumar M, Kumar P, Sinha AK, Anand U, Kumar A. Color Doppler-An effective tool for diagnosing midgut volvulus with malrotation. *Indian J Gastroenterol* 2017; 36: 27-31. <https://doi.org/10.1007/s12664-017-0729-5>
11. Drewett M, Johal N, Keys C, J Hall N, Burge D. The burden of excluding malrotation in term neonates with bile stained vomiting. *Pediatr Surg Int* 2016; 32: 483-486. <https://doi.org/10.1007/s00383-016-3877-2>
12. Goldman-Yassen AE, Gross J, Novak I, et al. Identification of clinical parameters to increase the diagnostic yield of the non-emergent upper gastrointestinal series in pediatric outpatients. *Pediatr Radiol* 2019; 49: 162-167. <https://doi.org/10.1007/s00247-018-4286-6>
13. Garcia AM, Asad I, Tessaro MO, et al. A multi-institutional case series with review of point-of-care ultrasound to diagnose malrotation and midgut volvulus in the pediatric emergency department. *Pediatr Emer Care* 2019; 35: 443-447. <https://doi.org/10.1097/PEC.0000000000001737>
14. Zhang W, Sun H, Luo F. The efficiency of sonography in diagnosing volvulus in neonates with suspected intestinal malrotation. *Medicine (Baltimore)* 2017; 96: e8287. <https://doi.org/10.1097/MD.00000000000008287>