

Assessment of developmental and radiological long-term outcome of children with surgically treated midgut volvulus

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Background/aim: The aim is to evaluate the long-term outcome of asymptomatic patients who underwent surgical correction for midgut volvulus.

Materials and methods: Seven patients managed surgically for midgut volvulus in the last 3 years were included. Demographic features, symptoms at presentation, diagnoses, surgical procedures, and complications were recorded. Patients were then contacted for follow-up and evaluation of long-term outcomes. General physical conditions, anthropometric parameters, feeding habits, and defecation histories were evaluated. Laboratory examinations were performed. Color Doppler ultrasonography (CDUS) was performed to evaluate blood flow in the superior mesenteric artery (SMA).

Results: In total, seven patients were identified. The median age at admission was 3 days (0–90 days). Mean age at follow-up was 17 ± 2 months. Growth parameters were normal in all cases. Four patients had low levels of ferritin and transferrin saturation. One patient had microcytic anemia. Another patient had low serum zinc level. One patient who had ileal resection had a high level of steatorrhea in stool examination. In CDUS, SMA blood flow volume was low in all cases. Peak-systolic velocity and resistance index were low in all but one case.

Conclusion: Despite uneventful postoperative courses, all patients operated on for midgut volvulus showed mild laboratory changes and decreased blood flow in the SMA in long-term follow-up.

Key words: Intestinal malrotation, intestinal volvulus, long-term, midgut, outcome

1. Introduction

All information about intestinal rotation and fixation anomalies accumulated over the years and became clear after the embryological description (1). ‘Malrotation or intestinal malrotation’ is a spectrum of disorders caused by abnormalities in intestinal rotation and fixation (1,2). These disorders range from typical malrotation to complete nonrotation, with an incidence of 1/500 live births. Midgut volvulus is seen in 6% of all malrotation cases (1,2).

Intestinal malrotation has a wide spectrum of symptoms and therefore leads to great variation in diagnosis and clinical presentation (1–3). Since midgut volvulus may cause a fatal clinical scenario, prompt diagnosis and early surgical management is vital in its

prognosis. Apart from the timing of surgical management, prognosis is also affected by postoperative complications such as intestinal adhesion, persistence of symptoms, sepsis, intestinal necrosis, and short bowel syndrome (4,5). However, there is a scarce amount of data regarding the outcomes of patients operated on for midgut volvulus.

In the present study, we aimed to evaluate the long-term outcome of asymptomatic patients who underwent surgical correction for midgut volvulus. Growth parameters and laboratory findings were used to evaluate the outcomes of patients. The role of blood flow in the superior mesenteric artery (SMA) in the postoperative outcome was evaluated by color Doppler ultrasonography (CDUS) in all patients.

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2. Materials and methods

The study was performed in adherence to the Declaration of Helsinki and with the approval of the local ethics committee (2014-20/01). Informed consent was obtained from the patients' parents before evaluation.

All patients who were surgically managed for midgut volvulus in the last 3 years were included in the study. The medical records of seven patients were reviewed retrospectively. The demographic features, symptoms, diagnoses, surgical procedures, and complications were recorded.

In the second part of the study, patients were contacted for follow-up and asked to come to our department for evaluation of long-term outcome based on general physical conditions, weights, heights, head circumferences, feeding habits, and defecation histories. Laboratory examinations including complete blood count, total protein and albumin levels, liver and renal function tests, lipid profiles, serum iron parameters, thyroid function tests, serum zinc level, urine analysis, and stool examinations were performed. All examinations were performed in an outpatient setting without hospitalization.

Additionally, all patients underwent CDUS examination of the SMA. All CDUS examinations were performed by the same radiologist using an Aplio scanner (Toshiba Medical Systems Co., Ltd., Tokyo, Japan) with a 5-MHz convex and 7.5-MHz linear-array transducer in a supine position after 6 to 8 h of fasting. All SMA measurements were performed 2–3 cm distal from its origin. Spectral Doppler analysis of the SMA was performed in the longitudinal section. During spectral analysis, the sampling gate was placed at the center of the vessel lumen and adjusted to encompass most of the lumen without touching the wall. The pulse repetition frequency was optimized to record medium to high arterial velocities in order to avoid aliasing. The Doppler angle of insonation was kept under 60°. We measured peak-systolic velocity (PSV), end-diastolic velocity (EDV), blood flow volume (BFV), and resistance index (RI) from spectral analysis of the SMA. Measured SMA BFVs were assessed according to the normal values that were calculated based on the formulas reported in the studies by Eloi et al. (6) [SMA blood flow = $256 + 329 \times (\text{body surface area}) \text{ mL/min}$] and Van Bel et al. (7) [SMA BFV = $43 \text{ mL/kg/min} \pm 13$]. Body surface areas (BSAs) were calculated as: $\text{BSA} = 0.024265 \times \text{height (cm)}^{0.3964} \times \text{weight (kg)}^{0.5378}$. PSV, EDV, RI, and PI values were assessed according to the normal values reported in the study by Okada et al. (8).

3. Results

Seven patients who underwent detwisting of intestines and Ladd's procedure were identified. Six of the infants were in the first week of their life and one infant was 3 months old.

The median age at admission was 3 days (0–90 days), and the male to female ratio was 3:4. The presenting symptom was bilious vomiting in all patients before the operation. Routine physical examination and biochemical analysis and blood counts were performed. Upright abdominal X-rays revealed dilated stomach, double bubble, and gasless abdomen appearance. Preoperatively CDUS was performed, revealing the classical 'whirlpool' sign, which is an indicator of intestinal volvulus. All patients were operated on immediately. Intraoperative findings included Ladd's bands and midgut volvulus in all cases. After detwisting of the intestines, necrosis was encountered in four cases and small bowel segments of 10–40 cm were resected and successfully anastomosed. Ladd's procedure was performed for all patients. The postoperative course was uneventful in all of them (Table 1).

The mean age at follow-up was 17 ± 2 months. All of the children's physical examinations were normal and all of them were asymptomatic. The weight values were in the 25th–50th percentile in 1 case, 50th–75th percentile in 5 cases, and 90th–95th percentile in 1 case. The height values were in the 25th–50th percentile in 1 case, 50th–75th percentile in 3 cases, and 75th–90th percentile in 3 cases. Feeding habits of all patients were normal. Only one was suffering from chronic constipation. None of them had short bowel findings including diarrhea or malabsorption.

The laboratory examination results, including complete blood count, liver and renal function tests, and lipid profiles, were within normal limits except for a low level of mean corpuscular volume in 4 cases. In four cases, ferritin and transferrin saturation levels were low. Another patient had microcytic anemia. These results showed that 5 of 7 patients had iron depletion, and in one of these 5 cases, iron depletion was at the level of iron deficiency anemia. Only one patient had a low serum zinc level ($65 \mu\text{g/dL}$; $N = 76\text{--}110$). Thyroid function tests were normal in all cases except for a slightly high TSH level in one case.

Urinalysis revealed moderated proteinuria in one case and a moderate level of uric acid crystals in another. There was high level of steatorrhea in the stool examination of one child who had a 20-cm ileal resection.

Values of SMA BFV were low in all patients when assessed according to the formulas given in the studies by Eloi et al. (6) and Van Bel et al. (7). PSV values, except in one patient, and RI values were also low according to the normal values given in the study by Okada et al. (8) EDV values of all patients were in normal limits. Measured Doppler parameters are given in Table 2. One of the patients (Case 4) was excluded from the BFV measurements since he had common SMA and celiac truncus.

Table 1. The summary of the medical history of the patients.

Case	Sex/age	Complaints	Radiological findings	Operation
1	F/3 days	Bilious vomiting	X-ray: air-fluid levels	Detwisting, Ladd procedure, resection-anastomosis
2	F/0 days	Bilious vomiting	X-ray: air-fluid levels	Detwisting, Ladd procedure, resection-anastomosis
3	F/8 days	Bilious vomiting	X-ray: air-fluid levels	Detwisting, Ladd procedure, resection-anastomosis
4	M/3 days	Bilious vomiting Bloody stool	X-ray: gasless abdomen	Detwisting, Ladd procedure
5	M/2 days	Bilious vomiting Abdominal distension	X-ray: double-bubble sign	Detwisting, Ladd procedure
6	M/3 months	Bilious vomiting	X-ray: air-fluid levels	Detwisting, Ladd procedure
7	F/0 days	Bilious vomiting Abdominal distension	X-ray: air-fluid levels	Detwisting, Ladd procedure, resection-anastomosis

Table 2. The growth parameters and color Doppler ultrasonography findings of the SMA.

Case	Age (months)	Weight (kg/percentile)	Height (cm/percentile)	BSA	SMA BFV	cSMA BFV*	cSMA BFV†	#PSV	#EDV	#RI
1	20	13.5/	86/	0.59	170	450	580 ± 13	100	16	0.84
		90-95	75-90							
2	9	10/	76/	0.47	290	410	430 ± 13	143	27	0.81
		50-75	75-90							
3	30	12.5/	89/	0.57	190	443	537 ± 13	135	16	0.85
		50-75	50-75							
4	24	13.4/50-75	86/50-75	-	-	-	-	-	-	-
5	24	12/	88/	0.54	320	433	516 ± 13	100	18	0.82
		25-50	25-50							
6	6	8.4/	70/	0.4	250	387	361 ± 13	102	16	0.83
		50-75	75-90							
7	17	9.8/	82/	0.4	260	387	421 ± 13	114	33	0.71
		50-75	50-75							

SMA: Superior mesenteric artery, BSA: body surface area, BFV: blood flow volume, RI: resistance index, PSV: peak-systolic velocity, EDV: end-diastolic velocity.

*: Values calculated from the SMA BFV formula = $256 + 329 \times \text{BSA}$ (6).

†: Values calculated from the SMA BFV formula = $\text{kg} \times 43 \pm 13$ (7).

#: Assessed according to the normal values given in the study by Okada et al. (8).

4. Discussion

Malrotation constitutes a wide spectrum of symptoms, diagnostic controversies, and fatal complications of

midgut volvulus. Not only short bowel syndrome but also postoperative problems continue to be frightening clinical scenarios for clinicians and surgeons (1-3,9,10). This wide

spectrum of presentation may lead to delayed diagnosis and fatal outcome. The initial diagnostic tool is mostly plain abdominal X-rays showing several specific signs such as double bubble or gasless abdomen. In suspected cases, upper gastrointestinal series and CDUS revealing a 'whirlpool' flow pattern of the superior mesenteric vein and mesentery around the superior mesenteric artery are further diagnostic tools (1,9,11). All patients in this study presented with bilious vomiting. One had bloody stool besides bilious vomiting. Only one of seven patients was 3 months old at presentation. Plain abdominal X-ray and CDUS were performed for our patients.

The standard management of midgut volvulus requires prompt diagnosis and early surgical management called Ladd's procedure should be performed immediately. Delayed diagnosis and management may lead to intestinal necrosis, wide segment bowel resection, short bowel syndrome, sepsis, and unfortunately death (4,12,13). Several complications may also be seen in patients who were diagnosed early such as intestinal adhesions, wound infections, and intestinal motility problems after Ladd's procedure (12,13). In addition to all these early postoperative catastrophes, malrotation may also cause long-term sequelae and complications. However, these long-term complications are less known, especially in asymptomatic cases in the postoperative period.

Although there are several studies regarding the outcome of patients with short bowel syndrome, there are not enough data about the outcome of asymptomatic patients operated on for midgut volvulus who had no short bowel syndrome. Murphy et al. presented long-term results after Ladd's procedure and revealed that three patients had malabsorption and needed dietary supplementation for 6 months (13). However, the outcome in clinically asymptomatic cases was not mentioned. El-Gohary et al. (4) stated that some patients had chronic constipation, gastroesophageal reflux, and recurrent abdominal pain, although most of the patients were incident-free at follow-up (4). In another study, constipation, failure to thrive, and abdominal pain were some of the complications seen in symptomatic patients after Ladd's procedure (5). However, none of the studies were able to show the correlation of symptoms with laboratory findings in both symptomatic and asymptomatic children. The clinical importance and consequences of monitoring patients without bowel

resection has also not been mentioned in most of the studies.

In the present study, all patients were asymptomatic and event-free. Their general physical condition and anthropometric parameters were all in normal ranges. Serum iron parameters revealed iron deficiency in four cases and zinc deficiency in one case. All patients were feeding well and only one case was suffering from chronic constipation. However, steatorrhea was detected in one patient who had a 20-cm bowel resection.

A literature review revealed that CDUS examination of the SMA has been used to determine Crohn disease activity or intestinal inflammation caused by other illnesses, bowel wall perfusion in preterm neonates to predict developing necrotizing enterocolitis, or in children with small bowel obstruction for the detection of strangulation (8,14,15). In the present study we intended to evaluate SMA blood flow hemodynamics after surgery and found low values of SMA BFV, PSV, and RI when assessed with the cut-off values given in the literature (6–8). We suggest that decreased blood flow in the SMA in patients with midgut volvulus may be related to congenital abnormality in anatomy such as a shortened mesenteric root and/or decreased intestinal length after resection. Additionally, there are not enough data as to whether decreased blood flow causes any clinical symptoms such as malabsorption or growth failure. Although it is difficult to draw a firm conclusion about the role of blood flow of the SMA in the etiology of postoperative problems, we suggest that decreased blood flow may be encountered in both symptomatic and asymptomatic patients. In order to identify the clinical importance of postoperative CDUS, randomized clinical trials comparing the blood flow parameters of the SMA in children with and without postoperative symptoms are needed. Additionally, future studies with larger numbers of patients are needed to confirm our results and define the clinical relevance of these findings.

In conclusion, patients operated on for midgut volvulus showed mild laboratory changes and decreased blood flow in CDUS evaluation of the SMA despite uneventful postoperative courses. It is hard to suggest long-term follow-up because of the unknown consequences of decreased blood flow in the SMA. Nevertheless, we think that this study may draw the attention of neonatal surgeons to evaluation of long-term outcomes of such patients.

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