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Original Article

Clinical Outcomes of Patients with Symptomatic Malrotation of the Intestine: A 25-year Experience

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Abstract

Objective: **The aim of this study was to review results of the treatment of patients with symptomatic malrotation within a 25-year period at a single tertiary institute in Thailand.**

Material and Method: **A retrospective study of patients with malrotation of the intestine who were treated at Queen Sirikit National Institute of Child Health between 1985 and 2009 was undertaken. Special attention was paid to clinical presentations, radiologic findings and results of treatment. Patients who were found to have malrotation as an incidental finding at laparotomy for other diseases were excluded from the study.**

Results: **A total of 153 patients underwent laparotomy for correction of symptomatic malrotation. Most of the patients developed symptoms, mostly bilious vomiting, within 30 days after birth. Upper gastrointestinal series were performed in 112 patients and revealed high gut obstruction, abnormal position of the duodenojejunal junction and corkscrew sign in 97 (87%), 74 (60%), and 38 (34%) patients, respectively. Patients were divided into three groups based on operative findings. Group A represented 58 patients, of whom 80% were neonates, who had only duodenal obstruction due to Ladd's band compression. Only one patient died postoperatively because of congestive heart failure from congenital heart disease. Group B included 75 patients who had midgut volvulus without intestinal necrosis. Approximately 70% of group B were neonates, and 3 died from complications of tetralogy of Fallot, severe pneumonia and gastric perforation, respectively. Group C included the 20 remaining patients who developed midgut volvulus with intestinal necrosis (involving 20% to 100% of the small intestine). Of these 20 patients, 17 (85%) were neonates and 8 (40%) died from extensive (> 70%) bowel gangrene. Patients who survived extensive bowel resection had the total length of viable small bowel over 30 cm. The mortality rate for all 153 patients was 7.8%.**

Conclusion: **Patients with malrotation developed symptoms within the neonatal period in approximately 80% of the cases. About two-thirds of intestinal malrotations presented with midgut volvulus and 20% of these had bowel gangrene. Necrosis over 70% of the small intestine was a significant risk factor for mortality in patients with midgut volvulus.**

Keywords: **Malrotation, midgut volvulus, intestinal necrosis**

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INTRODUCTION

Malrotation of the intestine usually presents with symptoms of intestinal obstruction in infancy and childhood, commonly caused by Ladd's band compression and midgut volvulus[1-4]. If symptoms first develop in older children, these may present with intermittent abdominal pain and vomiting and frequent repeated hospital admissions in some cases[5-7]. Intestinal necrosis secondary to midgut volvulus is the most serious complication of this abnormality[5]. Malrotation is sometimes found incidentally during surgical correction of abdominal wall defects, congenial diaphragmatic hernia and intestinal atresias. The authors undertook this retrospective review to determine the incidence of, and assess the impact of age at operation and eventual outcome in, patients with symptomatic malrotation of the intestine treated at a single tertiary institute over a 25-year period.

MATERIALS AND METHODS

After the study proposal (EC 142/2553) was approved by the Institutional Review Board, a retrospective review was performed of all patients with intestinal malrotation treated between January 1985 and December 2009. Information including clinical presentations, radiological findings, treatment and results were evaluated. Patients who were found to have malrotation as an incidental finding at laparotomy for other diseases such as omphalocele, gastroschisis, congenital diaphragmatic hernia, intrinsic duodenal obstruction, and other intestinal atresia or stenosis were excluded from the study. The data were analyzed by the Chi-square, Fisher's exact test, *t*-test, and Mann-Whitney U test as appropriate, with a p-value less than 0.05 considered significant.

Table 1 Clinical presentations in 153 patients

Symptoms / Signs	No. of patients	Percentage (%)
Bilious vomiting	119	78
Non - bilious vomiting	32	21
Abdominal pain	27	18
Abdominal distension	35	23
Hyperbilirubinemia	24	16
Bloody stool	16	11

RESULTS

During the 25-year period, 153 patients (111 males, 42 females) were operated on for symptomatic malrotation of the intestine at Queen Sirikit National Institute of Child Health. Eighteen of the 153 patients were born at Rajavithi Hospital, while the remaining were born at other hospitals. As the total number of neonates born at Rajavithi Hospital from 1985 to 2009 was 318,524, the crude incidence of symptomatic malrotation at Rajavithi Hospital was approximately 1 per 18,000 live births. The age of the patients ranged from one day to nine years. Approximately 80% (123 cases) were under one month old. Bilious vomiting was the most common presenting symptom, seen in 78% of patients (Table 1). Intermittent abdominal pain commonly developed in 27 older children (18%) and bloody stool was noted in 16 patients (11%).

Plain films of the abdomen revealed various

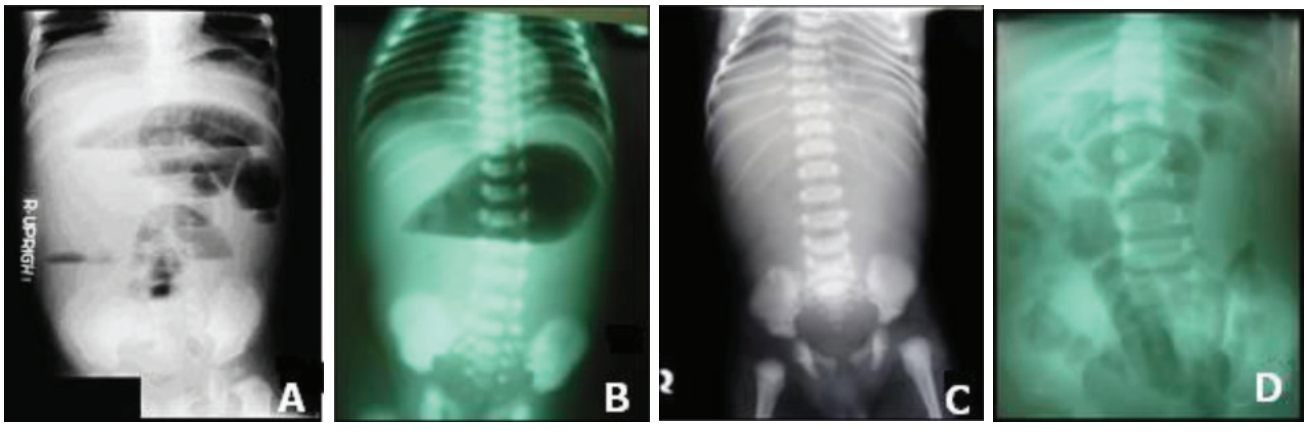

Figure 1 Plain films of abdomen in malrotation A. Complete small bowel obstruction B. High gut obstruction (only one gas bubble in the stomach) C. Gasless abdomen D. Normal finding

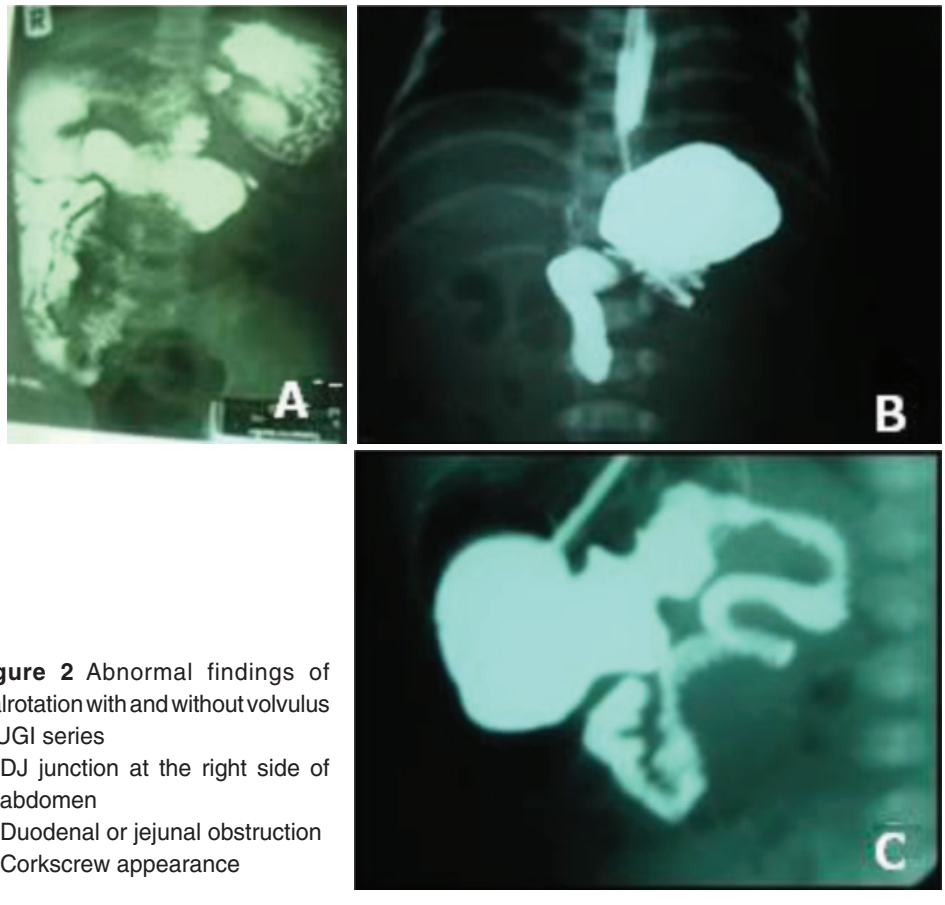

Figure 2 Abnormal findings of malrotation with and without volvulus in UGI series
A. DJ junction at the right side of abdomen
B. Duodenal or jejunal obstruction
C. Corkscrew appearance

patterns including small bowel obstruction, gas only in the stomach, gasless abdomen, and normal radiological finding in 115 (75%), 18 (12%), 15 (10%) and 5 (3%) patients, respectively (Figure 1). Upper gastrointestinal (UGI) series were done in 112 patients. Radiologic abnormalities included abnormal position of the duodenojejunal (DJ) junction, duodenal or jejunal obstruction and corkscrew appearance in 97 (87%), 74 (60%) and 38 (34%) patients, respectively (Figure 2). Barium enema (BE) was performed in only three cases and revealed abnormal position of the cecum in the right upper quadrant of abdomen (Figure 3).

All patients underwent laparotomy and were divided into three groups based on the operative findings. Duodenal obstruction, mostly at the second part, due to Ladd's band compression (group A) was noted in 58 patients (38%). The remaining 95 patients (62%) had malrotation with clockwise midgut volvulus (Figure 4). Seventy five of the 95 patients (79%) were found to have midgut volvulus with viable intestine after counterclockwise derotation of the volvulus (group B), whereas 20 patients (21%) with midgut

Figure 3 Abnormal position of the cecum at the right upper quadrant in a patient with malrotation

volvulus had intestinal necrosis (group C). Patients were operated on within the first month of life in approximately 80% of cases in group A, 70% in group B and 85% in group C; the median ages at operation were 10.5, 14, and 3 days, respectively. The average/

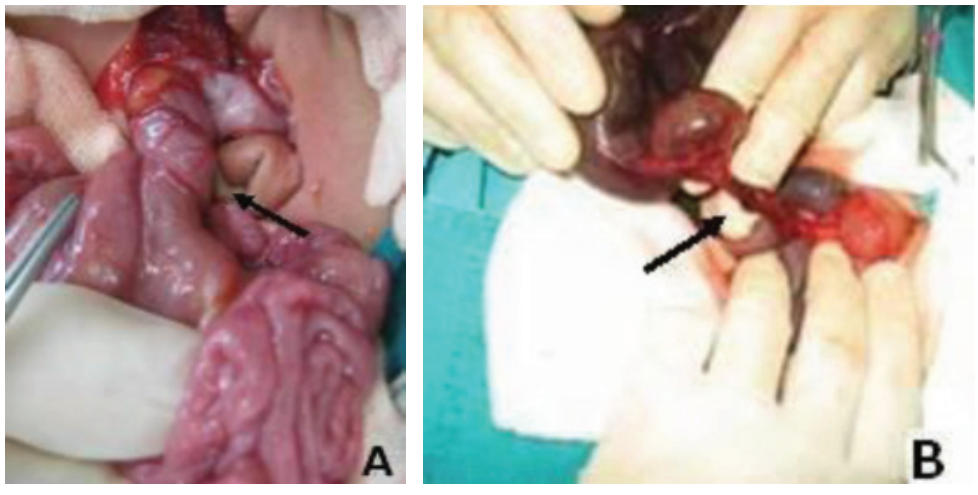

Figure 4 Presence of clockwise rotation in midgut volvulus
A. In a 25-day neonate without bowel necrosis
B. In a 8-day old neonate with extensive bowel necrosis

Table 2 Comparison of age operation and time interval from symptoms to operation between each group

Demographic data	Group A	Group B	Group C	p-value
Age at operation (days)				
Range	1-3285	3-3315	1-2035	A vs. B = 0.734
Mean	242.6±667.3	206.9±539.8	125.7±404.2	A vs. C = 0.464
Median	10.5	14	3	B vs. C = 0.532
Time interval from symptoms to operation				
Range	1-180	1-30	1-9	A vs. B = 0.100
Mean	13.7±28.7	7.9±8.0	2.7±2.3	A vs. C = 0.092
Median	4	5	2	B vs. C = 0.005

Table 3 Comparison of age at operation and mortality between each group

Age at operation	Group A		Group B		Group C	
	No.	Death	No.	Death	No.	Death
0 - 1 month	46	1	53	3	17	6
1 - 3 month	6	0	7	0	1	1
3 - 12 month	1	0	5	0	0	0
Over 1 year	5	0	10	0	2	1
Total	58	1 (1.7%)	75	3 (3.4%)	20	8 (40%)

Comparison between groups: group A vs. group B (1.7% vs. 3.4%, p = 0.631)
group A vs. group C (1.7% vs. 40%, p < 0.001)
group B vs. group C (3.4% vs. 40%, p < 0.001)

median age at operation of the patients in each group was not significantly different (Table 2). The average time interval from symptoms to operation in group A and group B was not significantly different (Table 2) but these were significantly or marginally higher than that of group C.

Regarding hospital mortality, 12 patients died (7.8%) and 10 of these (80%) were in neonatal period during presentation of symptoms and operations. Patients in group C had the highest mortality rate of 40%, whereas the mortality rates in group B and group A were 3.4% and 1.7%, respectively (Table 3). There was no recurrent midgut volvulus in the survivors. However, postoperative small bowel obstruction due to adhesions occurred in 5 of the 141 survivors (3.5%), 1 case in group A, 2 cases in group B and 2 cases in

Table 4 Associated congenital anomalies

Associated anomalies	No.	Associated anomalies	No.
Group A (27 cases)		**Group B (5 cases)**	
Congenital heart diseases	12	Anorectal malformations	2
Down's syndrome	5	Tetralogy of Fallot	1
Meckel's diverticulum	4	Situs inversus	1
Anorectal malformation	3	Dysplastic kidney	1
Situs inversus	2	Vesicoureteral reflux	1
Laryngomalacia	2	Mesenteric cyst	1
Omphalomesenteric duct	2		
Duplication of the duodenum	1	**Group C (2 cases)**	
Hiatal hernia	1	Anorectal malformation	1
Vertebral anomalies	1	Duplication of the jejunum	1
Other	3		

Some patients have more than one anomaly.

Table 5 Detailed description of the 20 patients in group C (midgut volvulus with bowel necrosis)

No	Age at operation (days)	Time from onset to operation (days)	Percentage of bowel necrosis (%)	Length of viable bowel (cm)	No. of operation	Major complications	Results
1	13	9	90	10	2	SBS[a], sepsis	dead
2	2	2	70	35	2	SBS[a]	survived
3	1	1	70	40	2	sepsis	dead
4	2	1	40	60	1	-	survived
5	8	2	40	60	1	-	survived
6	375	2	90	10	3	SBS[a], sepsis	dead
7	45	1	100[b]	0	2	sepsis	dead
8	2	1	80	25	2	anastomotic leak, sepsis	dead
9	2035	1	70	30	3	SBS[a]	survived
10	3	3	40	60	1	-	survived
11	5	5	40	65	1	-	survived
12	2	2	70	30	2	SBS[a]	survived
13	1	1	90	10	2	anastomotic leak, sepsis	dead
14	6	4	80	15	2	SBS[a], sepsis	dead
15	10	3	70	30	2	SBS[a]	survived
16	2	2	20	80	1	-	survived
17	3	2	90[c]	10	3	SBS[a], sepsis, gut obstruction	survived
18	2	1	100[b]	0	3	sepsis	dead
19	16	8	20	80	1	-	survived
20	3	2	60	40	2	SBS[a], sepsis, gut obstruction	survived

[a]SBS = short bowel syndrome; [b]No resection of necrotic bowel and no drainage; [c]Only Penrose drain placement, no resection of necrotic bowel

group C. Associated congenital anomalies were more common in patients in group A than those of other groups (Table 4). One patient in group A succumbed after Ladd's procedure and appendectomy due to severe congenital heart disease with congestive heart failure (CHF). Three patients in group B died postoperatively due to CHF from tetralogy of Fallot (1 case), severe pneumonia (1 case) and sepsis from preoperatively ruptured stomach (1 case).

The 20 patients in group C had midgut volvulus

with intestinal necrosis. Average ± SD and median percentage of small bowel necrosis was 66.5% ± 32.8% and 70%, respectively (ranged from 20% to 100%). All 20 patients underwent laparotomy with counter clockwise derotation of the volvulus and Ladd's procedure. A second look operation was noted in 10 cases (50%) and 4 cases (20%) required a third operation for definitive bowel resection. Only 6 patients (30%) had a segmental resection of necrotic bowel with appendectomy at the first laparotomy (Table 4). Eight patients (40%) succumbed from short bowel syndrome and septicemia. Of the 12 survivors 11 had small bowel necrosis ranging from 20% to 70% and the remaining viable small bowel length was at least 30 cm. They required extended treatment for short bowel syndrome. One exceptional case (No. 17) had small intestinal necrosis of approximately 90%, with remaining 10 cm of viable small intestine. The necrotic bowel was never resected. He developed many complications including enterocutancous fistula, septicemia, adhesive small bowel obstruction and prolonged paralytic ileus. He required four episodes of reoperation in a long hospital stay of over one year. He ultimately survived and was doing well in the last three years of follow-up.

DISCUSSION

The true incidence of intestinal malrotation is unknown because some cases are asymptomatic and remain undiagnosed. Bryne[8] reported an incidence of 1 per 6000 live births for symptomatic malrotation, compared with 1 per 18,000 live births in the present study. Estrada[9] estimated an incidence of 1 per 200 autopsies, and a clinical incidence of 1 per 25,000 admissions. Kantor[10] reported an incidental finding of malrotation in 0.2% of contrast studies of the gastrointestinal tract at any age. Many previous studies reported that 60% of the patients with malrotion developed clinical presentations within one month of age[1-3,11,12]. Our study revealed that approximately 80% of patients were under one month of age at surgery. Acute onset of bile-stained vomiting in neonates and infants with previously normal oral intake is the most common symptom of malrotation. Other manifestations included irritability or drowsiness and the passing of bloody stool. Chronic intermittent or vague abdominal pain with or without vomiting is commonly found in older children[5-7,11,12]. Passing of bloody stool was observed in 11% of our patients overall, but this presentation was noted in 80% of patients with midgut volvulus and intestinal necrosis (group C). Stewart[2] suggested that most neonates with obstruction from malrotation had association with midgut volvulus.

Plain abdominal radiographs revealed various findings such as small bowel obstruction, complete duodenal obstruction, only gas in the stomach, gasless abdomen or even normal intestinal gas distribution[11-14]. At our institute, UGI study is the investigation of choice for the diagnosis of malrotation with or without midgut volvulus. Classic radiological sign of malrotation is the malposition of the DJ junction at the right side of vertebral spine. A spiral or corkscrew appearance is the typical sign of midgut volvulus[3,11-14]. BE was historically recommended and is still used in some centers[14]. The advantage of BE is its relative safety in malrotation with midgut volvulus, but radiological findings may be difficult to interpret. BE may show abnormal position of the cecum, mostly in the right upper quadrant of abdomen. This finding may be found in some normal infants with incomplete fixation of the cecum at the right lower quadrant without malrotation. More recently color Doppler ultrasonography has been used to demonstrate the whirlpool sign for the diagnosis of midgut volvulus[15,16]. In addition, abdominal computerized tomographic (CT) scan can be also used to demonstrate superior mesenteric vein thrombosis in malrotation with chronic midgut volvulus[14].

Surgical correction by Ladd's procedure and appendectomy is the treatment of choice for intestinal malrotation. The entire bowel is mobilized from the peritoneal cavity to outside of the abdomen. Andrassy and Mahour[3] suggested that failure to extract the entire bowel outside the abdomen could lead to misdiagnosis or incomplete dissections of the bands. In patients with midgut volvulus, detorsion by counterclockwise rotation must be performed and the bowels should be covered with warm sponges to promote vascular recovery. If extensive intestinal necrosis is highly suspected, the abdomen should be closed and a second-look operation planned within 24-48 hours. We would like to suggest suturing only the skin or using of silastic sheath or other synthetic material to temporarily cover the abdominal wound in

order to prevent abdominal compartment syndrome during the wait for a second-look operation.

Many investigators have studied various factors which might influence hospital mortality, including age of patients[2,18,19] and delayed diagnosis[9,20]. The present study supported the association between mortality and younger age (Table 3). However, time interval between onset of symptoms and operation was not associated with an increase in mortality, especially if the obstruction or midgut volvulus was not tight. Messineo et al[21] studied clinical factors affecting mortality in children with malrotation of the intestine and found that significant risk factors included presenting symptoms in neonatal period, presence of serious abnormalities, and presence of necrotic bowel over 75%. These were similar to the findings of present study (Tables 3 to 5). Severe associated anomalies and preoperative complications were the major causes of death in patients with malrotation and midgut volvulus without bowel gangrene. Our study suggested that mortality was higher when the necrotic bowel was over 70%. Percentage of bowel necrosis did not correlate with degree of midgut volvulus and time interval from onset to operation. Extent of bowel necrosis was related rather to tight compression from clockwise volvulus of the midgut.

The incidence of postoperative adhesive small bowel obstruction was reported to vary widely from 3% to 24%[22,23]. This complication occurred in 5 of the 141 survivors (3.5%) in the present study. Murphy and Sparnon[24] emphasized long-term follow-up to parents regarding the risk and symptoms of postoperative small bowel obstruction in order to decrease the morbidity of this complication.

CONCLUSION

The present study demonstrated that clinical presentation of malrotation can occur at any age but approximately 80% occur within the neonatal period. Sudden bilious vomiting was the typical symptom in neonates and infants, whereas abdominal pain with or without vomiting was more common in older children. Two thirds of intestinal malrotation developed midgut volvulus and 20% of these had intestinal necrosis. Major causes of death in malrotion without intestinal necrosis included severe congenital anomalies and serious preoperative complications. For midgut volvulus with bowel gangrene, necrosis of over 70% of the intestine was an important risk for mortality. Patients who survived extensive bowel resection had viable small bowel of at least 30 cm.

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