

Increased risk of complications in acute onset intestinal malrotation

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ABSTRACT

INTRODUCTION: Intestinal malrotation is a potentially life-threatening illness which presents in many different ways and the symptoms span from acute to chronic. The purpose of this study was to determine the clinical presentation of intestinal malrotation at all ages.

MATERIAL AND METHODS: This was a retrospective study of all patients operated for intestinal malrotation at Odense University Hospital in the period from 1/1/1990 to 1/4/2012. The following information was recorded in the hospital records: demographics, presentation, duration of symptoms, diagnostic imaging and outcome. Patients were classified into two age groups: children (0-15 years) and adults (over 15 years).

RESULTS: A total of 54 patients were identified – 47 children and seven adults. Children frequently presented with acute symptoms, while adults mostly had chronic symptoms. The mortality rate was 6% and 14% for children and adults, respectively ($p = 0.44$). Deaths were due to volvulus.

CONCLUSION: Intestinal malrotation may have an acute and a chronic form in both children and adults. Complications and death only occurred in patients with an acute onset. Proper knowledge about symptoms is important for a good outcome.

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Intestinal malrotation is a relatively rare malformation with a reported incidence that varies from one per 6,000 to one per 200 live births depending on whether it includes symptomatic patients only or cases found by radiology or autopsy [1]. The term itself does not refer to a single congenital anomaly, but to a spectrum of development disorders in the normal rotation of the midgut.

Malrotation of the intestine occurs when the normal embryological rotation and fixation of the intestine does not occur [2, 3]. Intestinal rotation occurs during the second stage of the development of the midgut from 10-12 weeks of gestation. Midgut development is characterised by a strong longitudinal growth and a counter-clockwise rotation of 270° around the superior mesenteric artery.

The most serious complication is midgut volvulus, which is life-threatening and may lead to short-bowel syndrome if not operated within a few hours after acute

onset [3]. Other cases may present in a more chronic form with relapsing abdominal pain, nausea and vomiting. Finally, the condition may remain asymptomatic throughout life and be diagnosed as an occasional finding due to laparotomy for other diseases or at autopsy [4].

The purpose of the present study was to investigate symptomatic presentation, complications and outcome in patients operated for intestinal malrotation.

MATERIAL AND METHODS

This was a retrospective analysis of all patients operated for malrotation at Odense University Hospital during the period from 1/1/1990 to 1/4/2012. The material consisted of all records with the disease-specific coding for intestinal malrotation. The hospital is a tertiary referral centre for paediatric surgery covering approximately 2.5 million inhabitants and a primary referral centre for general surgery for approximately 325,000 inhabitants. Patients were divided into two age groups: children (0-15 years) and adults (over 15 years). A total of seven adults and 47 children were identified. From patient records, age, sex, characteristics of symptoms, symptom duration, radiological test, whether the correct diagnose was made before operation, complications to surgery and 30-day mortality after surgery were recorded.

The exclusion criteria were patients with no surgery and asymptomatic patients.

Statistical analysis

Summary statistics are presented as means \pm standard deviation. The groups are compared using Fisher's exact test for categorical variables, and p values below 0.05 were considered significant.

Trial registration: not relevant.

RESULTS

Demographics

Age at diagnosis varied from one day to 61 years. The adults had a mean age of 34.0 years ranging from 16 to 61 years. The children had a mean age of 237 days ranging from one day to 5.5 years. There was no statistically significant difference in gender between the two groups (Table 1).

ORIGINAL ARTICLE

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 TABLE 1

Demographics and symptoms.

	Children (N = 47)	Adults (N = 7)
<i>Demographics</i>		
Mean age, \pm SD	237 days \pm 495.4	34 yrs \pm 17.0
Male ^a , n (%)	31 (66)	3 (43)
Female ^a , n (%)	16 (34)	4 (57)
<i>Symptoms, n (%)</i>		
Acute onset	33 (70)	3 (43)
Pain	31	3
Bile vomit	29	2
Haematemesis/rectal bleeding	8	0
Chronic symptoms	14 (30)	4 (57)
Recurrent abdominal pain	14	4
Nausea/vomiting	12	4
Constipation/bloating	4	1
Diarrhoea	3	2
Weight loss	3	1
Gastroesophageal reflux	2	2

SD = standard deviation.

a) $p = 0.22$.
 TABLE 2

Duration of symptoms before diagnosis. The values are n (%).

	Children	Adults
<i>Acute onset</i>	33	3
Hours/days	27 (57)	1 (14)
Weeks	6 (13)	2 (29)
<i>Chronic onset</i>	14	4
Weeks	0	0
Months	12 (26)	0
Years	2 (4)	4 (57)

Twenty-seven of the 47 (57%) children were diagnosed within the first month of life. A total of eight (17%) children were diagnosed after the age of one year.

As our hospital serves as tertiary referral centre for paediatric surgery, it was possible to estimate the incidence of symptomatic intestinal malrotation in the Western part of Denmark (Funen and Jutland). In the period from 1990 to 2011, an average of 35,832 children were born each year [5]. In this period, a total of 47 were treated, which yields an incidence of 1:15,248.

Symptoms at presentation

Symptoms are presented in Table 1. 70% of the children presented with an acute onset. The most common symptom was pain (94%) followed by bile-stained vomiting (88%) haematemesis and/or rectal bleeding (24%). In all, 14 of the children (30%) had symptoms for more than a month with chronic or relapsing symptoms. The

most frequent symptom was abdominal pain (100%) followed by nausea/vomiting (90%). In the adult group, three (43%) had an acute onset with pain and 67% had bile-stained vomit. 57% of the adult patients had a history of recurrent abdominal pain and associated nausea and vomiting. Other symptoms appear from Table 1.

Table 2 shows the duration of the symptoms. There were no children or adults with symptoms lasting between 14 days and four weeks, and therefore no patients were classified as subacute. The children with acute onset mainly had symptoms from hours to several days, while adults with acute onset mainly had symptoms for more than seven days before a diagnosis was made or surgery performed. All adults with a chronic presentation had been having symptoms for years, and all children with a chronic presentation had symptoms for months.

Diagnostic methods

The majority (89% children, 86% adults) of patients had one or more diagnostic test performed before surgery (Table 3). The choice of method depended on the age of the patient. Children mainly had a plain abdominal X-ray and upper gastrointestinal contrast study (UGI). The adults mainly had a computed tomography (CT) or a UGI. Many of the children underwent more than one radiological examination. The one adult who did not have a radiographic test performed was pregnant.

UGI was used in three of five cases in adults who were diagnosed before surgery, and was diagnostic in all. The remaining two had a CT performed. In the children who were diagnosed preoperatively, 18 were diagnosed by UGI, two by barium enema and one by magnetic resonance imaging.

Operations and outcome

As presented in Table 4, all patients underwent surgery for malrotation. In the paediatric group, 37 (79%) were operated acutely, and 21% had elective surgery. In the adult group, 57% of the patients underwent acute surgery.

Complications included short-bowel syndrome, infections and ileus and occurred more frequently in the children.

DISCUSSION

The present study showed that there was an acute and a chronic presentation of malrotation in both children and adults. In children, acute onset was most common, whereas chronic symptoms were more common in adults. Patients with chronic symptoms presented with a wide variety of symptoms, which explained the diagnostic delay. Haematemesis and/or rectal bleeding, which must be considered as an important alarm symptom as



TABLE 3

	No imaging	Plain abdominal X-ray	UGI	Barium enema	CT	MRI	Diagnosis before surgery
<i>Children (N = 47)</i>							
Acute	5	18	12 (8)	2 (2)		0	10
Chronic	0	3	12 (10)	0		1 (1)	11
Total	5	21	24 (75%)	2 (100%)	0	1 (100%)	21 (45%)
<i>Adults (N = 7)</i>							
Acute	1	1	0		1 (1)		1
Chronic	0	0	3 (3)		1 (1)		4
Total	1	1 (0%)	3 (100%)	0	2 (100%)		5 (71%)

Diagnostic methods. The values are n (numbers in parentheses indicate the patients who were diagnosed before surgery, and percentages indicate the fraction of patients in which the investigation was diagnostic pre-operatively).

CT = computed tomography; MRI = magnetic resonance imaging; UGI = upper gastrointestinal contrast study.



TABLE 4

Operations and outcome. The values are n (%).

	Children (N = 47)	Adults (N = 7)
<i>Surgery</i>		
Acute surgery	37	4
Elective surgery	10	3
<i>Patients with complications</i>		
Short-bowel syndrome	2	1
Wound infection	5	0
Ileus	5	0
Reoperation	6	1
Mortality ^a	3 (6)	1 (14)

a) p = 0.44.

it might represent insidious intestinal gangrene, occurred in 24% of the children, but in none of the adults. Biliary emesis was the most common symptom in children, where it occurred in 58%, which is in accordance with previous publications [4]. Data regarding symptom descriptions were consistent with other studies [6, 7].

Two other studies comparing intestinal malrotation in all age groups have been published [4, 8]. One study included 170 patients and the other 24 patients. The study by Nehra & Goldstein [4] is surprising because it doubts that intestinal malrotation mostly presents during the first part of life. According to other literature, 80% of patients with malrotation are diagnosed during the first month of life, and 90% within the first year [3, 9]. In our study, the values were 56% and 84%, respectively. The incidence of intestinal malrotation is uncertain. International literature describes an incidence of one per 6,000 [1]. The incidence of malrotation was lower in our study than in other studies. However, our calculated incidence was based on the number of patients operated. The actual frequency of intestinal malrotation may be higher as the number of asymptomatic or fatal cases without surgery is unknown.

The cause of intestinal malrotation is unknown. Although it is not linked to a specific gene, it is most likely that a genetic component plays a role as some families have an increased incidence of malrotation [10].

The only treatment of symptomatic malrotation is surgery. Surgery includes rotating the intestine counter clockwise, placing the bowel in the most appropriate place, dividing the Ladd's band (the peritoneal band) and lysing any fibrous band or adhesions [9].

The diagnostic workout is challenging and our results do not allow any firm conclusions about the most appropriate investigation due to the relatively small number of patients within the different groups of investigation. The diagnostic procedure most used in children was plain abdominal X-ray and UGI, and the UGI had a higher diagnostic sensitivity. Similar findings have been reported by others [11]. Analysing UGI is difficult and requires an experienced paediatric radiologist. Abdominal X-ray is neither sensitive nor specific for malrotation. It can show bowel obstruction, but not intestinal malrotation. An alternative to UGI may be CT which is often preferred in adults [1, 4].

A significant number of the children with acute onset went directly to surgery without any investigation; immediate explorative laparotomy should always be performed in children suspected of suffering from volvulus as any delay of diagnostic investigations may be fatal due to intestinal necrosis.

Complications only occurred in acute surgery patients in both age groups, which explains the higher rate of complications in children due to a larger fraction presenting with an acute onset. The mortality rate was 6% and 14% for children and adults, respectively. The difference was not significant. Three deaths were caused by short-bowel syndrome. One death was caused by a post-operative ileus which led to intestinal necrosis.

CONCLUSION

There were two different types of presentation of mal-

Small bowel volvulus in a child with malrotation.



rotation. Acute onset was most common in children and recurrent symptoms were more common in adults. Among the diagnostic methods, UGI was the most efficient in diagnosing malrotation in children. Complications and death occurred only in patients with acute onset. Proper knowledge about symptoms is important for a good outcome.

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