

How Much of a Misnomer is “Asymptomatic” Intestinal Malrotation?

Zahavi Cohen MD, Oleg Kleiner MD, Robert Finaly MD, Jacob Mordehai MD, Nitza Newman MD, Edna Kurtzbart MD and Abraham J. Mares MD

Department of Pediatric Surgery, Soroka University Medical Center and Faculty of Health Sciences, Ben-Gurion University of the Negev, Beer Sheva, Israel

Key words: intestinal malrotation, midgut volvulus, “asymptomatic” malrotation

Abstract

Background: Intestinal malrotation is usually observed in the neonatal period with signs of acute high intestinal obstruction due to midgut volvulus. However, malrotation presenting beyond the neonatal period and well into adult life is associated with a variety of atypical and frequently non-specific gastrointestinal symptoms that may often cause prolonged delay in diagnosis and appropriate treatment.

Objectives: To emphasize the difficulty in predicting the risk of midgut volvulus based on age or symptoms, and to recommend surgery in all patients found to have intestinal malrotation even if they are considered asymptomatic.

Methods: We reviewed 41 patients with malrotation treated over a period of 24 years at the Soroka University Medical Center.

Results: In our series, 27 patients (66%) had acute midgut volvulus while 14 (34%) had malrotation found during investigation of various long-term gastrointestinal non-specific symptoms. Two patients died of total parenteral nutrition-related sepsis following extensive resection of small bowel. A total of 28 patients was available for long-term follow-up and are asymptomatic.

Conclusions: We recommend elective laparotomy and Ladd procedure in all patients found to have intestinal malrotation. This will prevent the catastrophic results of midgut volvulus and a variety of gastrointestinal symptoms wrongly attributed to other conditions in the span of a lifetime.

IMAJ 2003;5:172–174

Midgut volvulus is a dire surgical emergency in patients, mostly neonates, with intestinal malrotation. However, intestinal malrotation can present with a variety of less impressive, atypical and frequently non-specific gastrointestinal symptoms [1,2]. Such presentation, beyond the neonatal period, may elicit an erroneous interpretation and a delay in diagnosis and appropriate treatment. We describe our experience, emphasizing this entity and addressing the problem of “asymptomatic” malrotation.

Materials and Methods

We reviewed 41 patients with malrotation treated over a period of 24 years (1977–2000) in the Department of Pediatric Surgery at the Soroka University Medical Center. Excluded from this series were children with malrotation associated with omphalocele, gastroschisis, congenital diaphragmatic hernia, and intestinal atresias. Of the 20 males and 21 females, 24 (58%) were less than 1 month of age, 6 (14%) ranged in age between 1 month and 1 year, and 11 (28%) between 1 and 13 years.

Thirty-six patients underwent upper gastrointestinal contrast series that confirmed the diagnosis of intestinal malrotation with or without midgut volvulus. Five patients were operated for intestinal obstruction based on clinical symptoms, signs and abdominal plain X-ray. In all 41 patients, malrotation with or without midgut volvulus was diagnosed intraoperatively and Ladd procedure was performed. The operation included evisceration of bowel, inspection of the mesenteric root, devolvulization in a counter-clockwise fashion with assessment of bowel viability in cases of midgut volvulus, and intestinal resection whenever indicated. Also, division of all peritoneal bands, complete mobilization and straightening of the duodenum along the right abdominal gutter, appendectomy by inversion and placement of the cecum in the left lower quadrant were performed. Side-to-side duodenoduodenostomy was constructed in two patients with malrotation and preduodenal portal vein causing duodenal compression. One patient underwent, in addition, a Meckel's diverticulectomy. Bowel “pexy” was not done in our series.

Results

The patients were divided into three groups based on the clinical signs and symptoms and a history of previous gastrointestinal disorders.

- The first group of 18 patients (15 newborns and 3 infants under 1 year old) presented clinically with signs of acute abdomen and/or high intestinal obstruction. All had midgut volvulus at operation. None had any gastrointestinal disorders before the acute onset of symptoms.
- The second group of 9 patients (3 newborns, one 4 month old infant, and 5 children aged 1–13 years) also presented with acute signs of intestinal obstruction and midgut volvulus at surgery, but they were characterized as having had a history of recurrent gastrointestinal disorders. The three newborns (aged 19, 23 and 27 days) had recurrent vomiting, feeding problems, diarrhea, and failure to thrive from birth. They were all initially diagnosed as suffering from milk allergy. Six children in this group were hospitalized once or more prior to their definitive diagnosis. These children suffered from various gastrointestinal symptoms for a period ranging from 1 week to a few years.
- The third group of 14 patients was characterized by a long-standing history of a variety of gastrointestinal symptoms. Some of them were hospitalized at least once because of episodic attacks of abdominal pain, vomiting and diarrhea, mostly

diagnosed as milk allergy or gastroenteritis. Eventually, they had an upper GI contrast series and were finally diagnosed as having intestinal malrotation. They were electively operated upon, the diagnosis was confirmed and, in addition, all had signs of chronic "on and off" volvulization, i.e., thickened mesenterium, lymphatic and venous congestion, mesenteric lymph node enlargement, and various amounts of intraperitoneal chyle.

Three patients (from the first and second group) with midgut volvulus had varying degrees of intestinal necrosis necessitating small bowel resection of various length.

Postoperative complications developed in seven patients (17%): wound infection in two, adhesive small bowel obstruction in one, short gut syndrome in two, prolonged atony of duodenum in one and seizures in one. Two patients died at the age of 3 and 3.5 months respectively. Both patients had extensive resection of necrotic small bowel and their death was due to total parenteral nutrition-related sepsis.

There were no cases of recurrent volvulus in our series. Twenty-eight patients were available for follow-up from 1 to 15 years following surgery. Three patients suffered from periodic vomiting, one of whom also had periodic diarrhea following surgery. All three patients were neonates at operation and these symptoms subsided between the ages of 7 and 13 months. All the rest were thriving and asymptomatic.

Discussion

Midgut volvulus is the most serious complication of intestinal malrotation, hence, early recognition and surgical intervention are mandatory. It may occur *in utero*, causing variable degrees of ischemic necrosis and resulting in a single or multiple atresias of the small bowel [3]. However, it is usually observed in the neonatal period with signs of an acute high intestinal obstruction due to midgut volvulus or Ladd's bands that partially obstruct the duodenum [4,5]. In our series, 58% of the patients were newborns presenting with a short history of bilious vomiting. Beyond the neonatal period, acute presentation with midgut volvulus, although possible, is less frequent. In our series we found nine such patients (22%).

The important group that we would like to emphasize is patients with histories of non-specific gastrointestinal disorders such as intermittent abdominal pain and/or vomiting, feeding problems, or recurrent diarrhea. These patients are often diagnosed as suffering from milk allergy, malabsorption, celiac syndrome, or even psychological disorders [1,4,6]. In our series, 23 patients (56%) fit this category, of whom 9 eventually developed midgut volvulus and were operated urgently. The rest, 14 patients, did not develop midgut volvulus and were finally diagnosed by upper GI series as having intestinal malrotation and were operated electively.

This experience emphasizes the difficulty in reading the precise diagnosis in a variety of patients with non-specific gastrointestinal symptoms. Intestinal malrotation can be an elusive state of affairs. Even when "incidentally" diagnosed, usually by an upper GI series,

there is always a cause for alarm. It is hard to accept the term "asymptomatic" malrotation, as some authors have labeled this entity [7,8].

Management of the so-called asymptomatic patients with malrotation is controversial. Some authors believe that observation is appropriate for older children and advocate selective surgical treatment according to specific anatomy based on upper GI contrast studies [9,10]. Others, like von Flue et al. [11], are convinced that malrotation always requires surgical treatment. In our opinion, "asymptomatic" intestinal malrotation is a misnomer in the majority of cases. We believe that real asymptomatic patients are those in whom malrotation was an incidental finding at laparotomy for other conditions, or discovered at autopsy and even these patients' histories should be scrutinized thoroughly for hints of previous gastrointestinal symptoms. No patients in our series were asymptomatic. All of them had protracted gastrointestinal disorders that subsided following surgery.

Some authors emphasize that patients presenting with symptoms of malrotation during the first month of life are at risk of developing midgut volvulus [12]. Others reported that the majority of patients with volvulus were less than 1 year of age [13]. The occurrence of midgut volvulus in infants presenting in the neonatal period ranged from 45 to 80%, while the frequency among older patients has been reported as low as 14% [14]. In our series, among the 27 patients with midgut volvulus, 18 (66%) were neonates while 34% were infants and children under 13 years old. Although the risk of developing midgut volvulus is definitely highest in the neonatal and infancy group in most series, it can occur at all ages, even through adult life [15]. Spigland et al. [2] reported an almost equal incidence of midgut volvulus among both age groups (65% versus 85%). Wang and Welch [16] reported that 50% of adults reviewed in large series developed symptoms related to their malrotation. Von Flue et al. [11] reported a series of 10 adults with malrotation, whose age ranged from 21 to 72 years. In their series, four patients presented acutely – one with midgut volvulus and three with ileocecal volvulus. The other six patients had chronic gastrointestinal symptoms. All patients underwent the Ladd procedure and were symptom-free 2 to 10 years following surgery.

Based on our experience as well as others, we recommend the following:

- A high index of suspicion in young children with "vague" recurrent gastrointestinal symptoms, often labeled as "milk allergy"
- The labeling of any proven malrotation as "asymptomatic" is dangerous, in view of the possible occurrence of midgut volvulus throughout a life-span often without any early warning
- Surgical correction of documented malrotation should be carried out in all patients, regardless of age.

References

1. Kullendorf CM, Mikaelsson C, Ivancev K. Malrotation in children with symptoms of gastrointestinal allergy and psychosomatic abdominal pain. *Acta Paediatr Scand* 1985;74:296–9.
2. Spigland N, Brandt ML, Yazbeck S. Malrotation presenting beyond the neonatal period. *J Pediatr Surg* 1990;25:1139–42.

GI = gastrointestinal

3. Nixon HH, Tawes R. Etiology and treatment of small intestinal atresia: analysis of a series of 127 jejunal atresias and comparison with 62 duodenal atresias. *Surgery* 1971;69:41–6.
4. Millar AJW, Rode H, Brown RA, Cywes S. The deadly vomit: malrotation and midgut volvulus. *Pediatr Surg Int* 1987;2:172–6.
5. Seashore JH, Touloukian RJ. Midgut volvulus. An ever-present threat. *Arch Pediatr Adolesc Med* 1994;148:43–6.
6. El-Gohari JH, Cook RCM. Intestinal malrotation beyond the neonatal period. *Z Kinder Chir* 1984;39:237–41.
7. Gohl ML, De Meester TR. Midgut nonrotation in adults and aggressive approach. *Am J Surg* 1975;129:319–23.
8. Silverman A, Roy C. Pediatric Clinical Gastroenterology. St Louis: Mosby, 1983:62–6.
9. Filston HC, Kirks DR. Malrotation – the ubiquitous anomaly. *J Pediatr Surg* 1981;16:614–20.
10. Schey WL, Donaldson JS, Sty JP. Malrotation of bowel: variable patterns with different surgical consideration. *J Pediatr Surg* 1993;28:96–101.
11. Von Flue M, Herzog U, Ackerman C, Tondelli P, Harder F. Acute and chronic presentation of intestinal malrotation in adults. *Dis Colon Rectum* 1994;37:192–8.
12. Prasil P, Flageole H, Shaw KS, Nguyen LT, Youssef S, Laberge JM. Should malrotation in children be treated differently according to age? *J Pediatr Surg* 2000;35:756–8.
13. Stewart DR, Colodny AL, Daggett WC. Malrotation of the bowel in infants and children: a 15 years review. *Surgery* 1976;79:716–20.
14. Fukuda T, Brown BP, Lu CC. Midgut volvulus as a complication of intestinal malrotation in adults. *Dig Dis Sci* 1993;38:438–41.
15. Berardi RS. Anomalies of midgut rotation in the adult. *Surg Gynecol Obstet* 1980;151:113–24.
16. Wang CA, Welch CE. Anomalies of intestinal rotation in adolescents and adults. *Surgery* 1963;54:839–55.

Correspondence: Dr. Z. Cohen, Dept. of Pediatric Surgery, Soroka University Medical Center, P.O. Box 151, Beer Sheva 84101, Israel.
 Fax: (972-8) 623-2305
 email: zahavic@bgu.mail.bgu.co.il

Research Projects

Psychological and physical health among family caregivers of the chronically ill: a comparison study of immigrant and veteran Israeli residents

V. Soskolne PhD

School of Social Work, Bar-Ilan University, Ramat Gan, and Department of Social Medicine, Hadassah Medical Organization, Jerusalem, Israel

Background: Family caregiving for the chronically ill is a stressful situation that is often associated with adverse psychological and physical health outcomes. Immigrant caregivers may be more vulnerable. The study examines cultural/immigration status differences based on current views of the coexistence of positive as well as negative aspects of psychological health outcomes in the stress and coping model.

Objectives: To compare the caregiving stressors and health (psychological distress, negative and positive affect, physical health) between recent immigrant caregivers from the former Soviet Union and veteran resident caregivers, and to examine the contribution of caregiving stressors and psychosocial variables to the differences in health.

Methods: A total of 207 immigrant and 212 veteran resident caregivers, recruited from all the home-care units in Jerusalem and three ambulatory units at Hadassah Medical Center, were interviewed.

Results: The immigrant caregivers reported higher levels of caregiving activities (stressors). The differences in psychological health varied by relationship to care recipient. The immigrant spouses reported *lower* levels of negative affect ($P<0.01$) but no other differences, while the immigrant adult children reported *higher* levels of psychological distress and *lower* positive affect ($P=0.05$). Physical health of the immigrant caregivers was significantly worse irrespective of relationship type. In multivariate analyses, caregiving stressors and psychosocial variables mediated the association between immigration status and psychological health variables, but not the association with self-assessed health.

Conclusions: Immigrant adult child caregivers should be targeted for intervention to decrease their greater risk for elevated distress and lower positive affect by enhancement of psychosocial variables. The worse physical health status of the immigrants is independent of their caregiver stress.

Supported by the Office of the Chief Scientist, Ministry of Health, Jerusalem