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

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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 parental 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.

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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, devascularization 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 symp-
toms 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 90% 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"
recurring 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

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GI = gastrointestinal

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**Research Projects**

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

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**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 German 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 some 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.

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