

# MIDGUT MALROTATION IN OLDER CHILDREN

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**Abstract-** Midgut malrotation is typically presented during the first few months of life but sometimes may encounter later in life, causing difficulties and mistakes in diagnosis. We reviewed records of eleven rare patients with midgut malrotation older than one year of age and extracted their clinical and paraclinical data. The most common presenting symptoms were bilious vomiting, recurrent abdominal pain and constipation. Five of eleven patients had presented from neonatal period. The average interval between first symptoms and surgical correction of malrotation was about 22 months. Some of the patients had been undergone false treatments. Most cases were diagnosed by contrast studies (upper gastrointestinal series and barium enema). Diagnosing midgut malrotation in older children is often delayed. This anomaly should be suspected in all children with signs and symptoms of small bowel obstruction, chronic abdominal pain and vague abdominal discomfort and in all patients of any age with abdominal discomfort who do not respond to other therapies. Contrast studies may be necessary to rule out malrotation in suspected patients.

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**Key words:** Midgut malrotation, diagnosis and children

## INTRODUCTION

Intestinal or midgut malrotation (MM) is an anomaly with broad spectrum of clinical manifestations. The common presentation is upper intestinal obstruction usually in the first year of life especially in neonatal period (1, 2) but there are reports of rare cases with delayed presentation in late adulthood (3, 4). The clinical presentation of MM in older children and adults is usually nonspecific with a history of episodic abdominal pain and vomiting (5, 6) and are usually due to chronic complications of MM including chronic duodenal obstruction, chronic midgut volvulus and internal herniation. In this article, we reviewed 11 children with midgut malrotation who were older than one year of age.

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## MATERIALS AND METHODS

We reviewed clinical records from 57 patients with an established diagnosis of intestinal malrotation in the "Children Medical Center", a tertiary care center in Tehran University of Medical Sciences from January 1983 to September 2005.

Diagnosis was established on clinical grounds and surgical findings. We excluded patients with other anomalies such as abdominal wall and diaphragmatic hernias that interfered with the presentation and symptoms. Among 46 patients with final diagnosis of intestinal malrotation there were 11 children who were treated after first year of life by surgical Ladd's procedure.

The information about clinical presentation, evaluations, misdiagnoses and delays in diagnosis and treatment was collected.

## RESULTS

We reviewed 11 patients with a median age at the time of diagnosis of 28 months (range: 1 year to 11 years) and nine patients were male (82%). The most

## Midgut malrotation in older children

common symptoms were bilious vomiting (100%), recurrent abdominal pain (54%) and constipation (45%). Other symptoms and signs were dehydration (36%), abdominal distention (18%), fever (18%), weight loss (18%), obstipation (18%) and diarrhea (18%). Median age of first presenting symptom was 30 days. The average interval between first symptoms and surgical correction of malrotation was about 22 months. Mean interval from admission to surgery was 7.3 days (Table 1).

Previous diagnoses labeled to these patients before definite diagnosis of malrotation are listed in table 2. Some patients had been treated for a long time with wrong diagnosis: A five-year-old boy with intermittent abdominal pain with diagnosis of abdominal epilepsy treated with carbamazepine and metoclopramide for a year. He had been referred as a case of resistant abdominal epilepsy. Another case was an 11-year-old boy presented with 6-month history of chronic abdominal pain radiated to his back and relieved with flexed legs into the abdomen or in knee-chest position. He had been assessed with suspicion to pancreatitis. He had been undergone many investigations including five abdominal ultrasonographies, upper gastrointestinal endoscopy and gastric biopsy, endoscopic retrograde cholangiopancreatography (ERCP) and upper gastrointestinal contrast study. Serum amylase and sweat test had been checked for several times that all were normal. He had been treated with antibiotics, somatostatin, cisapride, hyoscine and undergone courses of total parenteral nutrition.

Diagnostic evaluations of these patients are listed in table 3.

**Table 1.** Chronology of presentation and treatment of older children with midgut malrotation

<b>Age at first symptom presented</b>	
Median	30 days
Mean $\pm$ SD	765 $\pm$ 1240 days
<b>From first symptom to diagnosis</b>	
Median	465 days
Mean $\pm$ SD	658 $\pm$ 754 days
<b>Interval from admission to surgery</b>	
Median	6 days
Mean $\pm$ SD	7.3 $\pm$ 7.3 days

**Table 2.** Differential Diagnoses of patients before definite diagnosis of malrotation

Intussusception
Acute Appendicitis
Crohn disease
Ulcerative Colitis
Gastroesophageal Reflux *
Gastritis *
Peptic disease *
Irritable Bowel Syndrome *
Abdominal epilepsy *
Abdominal migraine
Pancreatitis *
Cystic Fibrosis
Malabsorption Syndrome
Functional Constipation *
Lymphoma
Tuberculosis
Parasitic infection *
Necrotizing Enterocolitis

\* Wrongly treated with this diagnosis.

In our series, malrotation had been diagnosed more often by contrast studies (upper gastrointestinal series and barium enema). None of our five patients evaluated with ultrasonography were diagnosed by this modality. Complications of malrotation occurred in one case. She was a 6.5-year-old girl with intermittent vomiting and constipation after birth and several admissions and a laparotomy in neonatal period that only lymph node biopsy had been done. She presented with acute abdomen due to superior mesenteric vein thrombosis and extensive small bowel necrosis. There was no mortality in our patients.

**Table 3.** Diagnostic evaluations and their results

<b>Diagnostic evaluation</b>	<b>No. of</b>	<b>No. of cases</b>
	<b>Cases</b>	<b>Diagnosed</b>
	<b>evaluated</b>	<b>(%)</b>
Plain abdominal X-Ray	7	4 (57%) *
Upper GI Series	7	5 (71%)
Abdominal Sonography	5	0 (0%)
Barium Enema	3	3 (100%)
Upper GI Endoscopy	3	1 (33%) <sup>S</sup>

\* In 2 diagnosed Duodenal Obstruction and in 2 diagnosed GI Obstruction

<sup>S</sup> Suspicious to malrotation

## DISCUSSION

Symptoms of midgut malrotation in older children and adults are obscure and usually of prolonged duration (7, 8) and sometimes atypical (9, 10). This leads to misleading the physician to other diagnoses and delay in diagnosing the problem (11). In a series of 51 patients under the age of 10 with malrotation, this was the primary diagnosis in only 20 cases (39%) (12). As shown in our review, the mean time from first symptom to correct diagnosis was 1.8 years with a maximum of 6.5 years. Our review in patients' records showed that nearly half of them (45%) had symptoms from neonatal period and missing the diagnosis occurred mostly in the first months of life; then the older became the patient, the less probable a physician became suspicious to MM. The remaining half had delayed presentation.

Complications of intestinal malrotation, mainly midgut volvulus, can occur in any age and even adult patients. Prompt recognition and surgical treatment usually lead to a successful outcome but there is considerable morbidity and mortality in patients with intestinal necrosis. Another unusual complication of malrotation with chronic midgut volvulus that has been reported is superior mesenteric vein thrombosis (13), as in one of our cases. These catastrophic complications emphasize the need for early detection and consideration of Ladd's procedure for patients of all ages because it remains impossible to predict which patients will have complications. Therefore, even incidentally discovered patients with intestinal malrotation should undergo Ladd's procedure (5, 14).

Several methods can be used to diagnose MM. Our patients were mostly diagnosed by contrast studies and ultrasonography helped in none of them. Powell reported 70 patients with malrotation that upper gastrointestinal series revealed the diagnosis in 29 cases (41%), as did contrast enema in 24 (34%) (15). Modern ultrasound examination is also called helpful (16, 17). This method is proposed as a screening test but in our review, ultrasonography was incapable of diagnostic value, maybe because of lack of experience of sonographers with sonographic patterns of malrotation in last decades and also because most of sonographic evaluations were done

with less suspicion to MM. Therefore it is recommended to use contrast studies in suspected cases. If ultrasonographic evaluation is to be done, the suspected diagnosis should be discussed with sonographer.

"Not considering MM as a cause of intestinal disorder after first months of life" seems to be a major factor of delay in detecting this anomaly in older children. Therefore, these patients may be mistreated for a long time. A high index of suspicion is required in evaluating children with possible malrotation and it must be discussed with other medical staff like radiologists. The diagnosis of malrotation should be considered in any child and even adult with intermittent abdominal pain, vague abdominal discomfort and gastrointestinal symptoms, especially in patients with other diagnoses who do not respond to their treatments. Misdiagnosis is usually followed by unnecessary assessments and incorrect treatments. Review of our patients showed that the most unnecessary studies have been done outside of surgery ward. Any suspected patient must be consulted with a pediatric surgeon.

Patients with misdiagnoses suffer from their abdominal problems as well as multiple admissions and also are predisposed to "Acute Midgut Volvulus" that is tremendous and life-threatening. It is important to note that volvulus, intestinal gangrene, and mortality occurred regardless of age or chronicity of symptoms (15).

### Conflicts of interest

The authors declare that they have no competing interests.

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## Midgut malrotation in older children

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