Presentation of Intestinal Malrotation Syndromes in Older Children and Adults: Report of Three Cases
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Malrotation syndromes are typically presented during the first few months of life but may sometimes appear later in life, causing difficulties and mistakes in diagnosis. We had 40 cases of malrotation, 3 of them with atypical and delayed presentation. Two of the three cases had a history of abdominal disorders that lasted for several years. Severe dehydration was a significant symptom in one of them. Diagnosis was delayed because respective symptoms were not adequately considered in older children and adults.

Key words: duodenal obstruction; ileus; intestinal obstructions; volvulus

Many individuals with intestinal malrotation never develop the syndrome in the older age. Patients who were previously treated for congenital diaphragmatic hernia or congenital abdominal wall defects have intestinal malrotation as an associated anomaly (1). In most of the patients followed up after Ladd procedure due to intestinal malrotation in infancy there was no recurrence of the symptoms (2). Ladd procedure stabilizes but does not remove the anomaly. Also, there are people who are not aware that they have the anomaly. Minor or severe abdominal disorders may be present for a long time or the anomaly may be completely asymptomatic.

Malrotation syndromes are clinically evident in the first month of life in 64% of the patients with the syndrome and in 82% in the first year of life (3,4). The onset of clinical presentation is rare later in life (5,6) and the symptoms are obscure and usually of prolonged duration (7). The diagnosis is often delayed and various others, including psychogenic disorders, are made before a final, correct one (8).

Bill divided intestinal malrotation according the time of occurrence in three embryonic stages (9).

Malrotation syndromes in children are most commonly presented as Ia or IIIa type. These are also the most serious types. Clinically, they are manifested as duodenal obstruction caused by volvulus of the midgut (10,11). They become apparent when the gut is filled with air in the first days of life. If a complete volvulus does not develop, the patients present with the symptoms of subtotal obstruction of the duodenum and the ileus develops later. Other types are very rare and are often found accidentally at operations performed for other reasons. After the second year of life, the manifestations of malrotation are very rare. These patients develop stable subtotal obstruction of the duodenum and/or intermittent volvulus (12-14). The prevalence of nonrotation without volvulus and symptoms of obstruction is not known (9). During the period of 10 years (January 1, 1986 - December 31, 1995), 3 out of 40 of our surgically treated patients with intestinal malrotation had a delayed presentation.

Case 1
E.A., a 7 year-old female child failed to normally grow after birth. She did not eat meat, fruit, and vegetables, and did not drink milk because of vomiting after each meal. Vomiting was also the first symptom of common childhood infections (tonsillitis, uroinfection). She was intensely vomiting for three days prior to the admission and developed symptoms of dehydration. On admission, the abdominal wall was soft and generally slightly tender. Peristalsis was weak. Abdominal X-ray showed an air level in the duodenal area. Giant, dilated stomach was shown by contrast examination, as well as the obstruction of the horizontal part of the duodenum. On surgery, nonrotation of the Ia type with volvulus of the midgut was found. There were no ischemic changes of the gut and Ladd procedure (15-17) was performed. Postoperatively, the clinical status improved and normalized without any complications.

Case 2
T.R., a 15 year-old male child, had been complaining of abdominal pain around the umbilicus for years, and had been previously diagnosed and treated for a pancreatic cyst. He was vomiting for four days prior to admission. On admission, the pain was localized in the ileocecal region. An operation was indicated for suspected appendicitis. Appendical inflammation was not found and the abdominal cavity was closed after appendectomy. After the operation, vomiting continued, as well as a colic type of pain. A “double-bubble” sign on X-ray was found. Gastroduodenal contrast examination revealed
an extremely dilated stomach and bulbous duodenum, as well as an obstruction of the horizontal part of the duodenum. Malrotation of the Ia type (nonrotation) was found during surgical exploration and the Ladd procedure was performed. The patient is at present completely symptom-free.

Case 3
Š.Z., a 32 year-old man, presented with symptoms of acute abdomen. He did not have any abdominal disorders before. Twelve hours before admission, he felt a strong colic pain under the right rib arch and began to vomit. Clinically, his abdomen was slightly distended and extremely painful in the right lower quadrant. No peristalsis but gurgling water phenomena were heard. Giant air level in the right abdomen was found on X-ray examination. The site of obstruction was in the area of the cecum and ascendent colon was visualized by a barium enema (bird beak sign) (18). During surgery, malrotation of the IIIc type and volvulus of the cecum were found. After a detorsion maneuver, the compromised circulation improved, and cecopexy to the lateral abdominal wall was performed. There were no complications after surgery.

Discussion
The reason for a delayed presentation of malrotation syndromes is not known. There is a balance between the narrowing of the duodenum and the torsion of the midgut. The degree of the duodenal obstruction may influence clinical symptoms. It may be obstructed enough to cause some disorders, but not enough to cause ileus. The syndrome appears at the point of critical narrowing (9,19,20) This implies that many people live with asymptomatic intestinal malrotation or have only small disorders. Two of our patients had abdominal problems presented as abdominal pain and vomiting for many years. In the differential diagnosis in Case 2, malrotation syndrome was never considered. When duodenal ileus developed, it resulted in a diagnostic and surgical mistake. Delayed diagnosis of intestinal malrotation may be responsible for severe dehydration (21) as in our Case 1. In both cases the type of malrotation was nonrotation.

Today, most of the cases are diagnosed before the acute onset of the disease (22). Modern ultrasound examination is very helpful in the diagnosis. The position of the superior mesenteric vein left to the artery suggests malrotation in almost all cases. The position of the vein anterior to the artery is found in 30% cases of intestinal malrotation (23,24). This method is applicable as a screening test when considering a differential diagnosis.

The most common type of malrotation is IIIc. It can be found in 10-19% of all population. It is mainly asymptomatic but causes differential diagnosis and technical difficulties in the surgical treatment of appendicitis. It is rarely presented as a volvulus of the cecum, mostly in adults (25). The absence of medial (valvula Bauchini) and lateral (“gutter”) fixation of the cecum make possible its axial rotation. Volvulus probably appears when the cecum surpasses a critical size. This may be the reason why it is rare in children. The interesting association between intussusception and intestinal malrotation (26), where the malfixation of the cecum can be important, requires further consideration.

None of our patients treated for and surviving congenital diaphragmatic hernia, gastro- schisis or omphalocele accompanied with intestinal malrotation ever developed malrotation syndrome. However, some of them had transient, even severe abdominal disorders due to the short gut syndrome (absorption), intestinal neurogenic immaturity (motility) or intestinal malrotation itself (obstruction?).

In our experience, there was no recurrence of symptoms after the Ladd procedure, although they have been described (1,27). We also did not encounter adhesive ileus either as duodenal or vascular obstruction. In the long term follow-up, there were only minor abdominal disorders of transient abdominal distension and/or vomiting, non-specific pain or constipation. In conclusion, intestinal malrotation should be considered as a differential possibility in the diagnosis of abdominal disorders in older children and adults. Using ultrasound examination as a screening test, the anomaly could be observed before the syndrome develops.

References

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