

# Obsežna resekcija tankega črevesa pri malrotaciji z akutnim volvulusom pri tri letnem dečku: prikaz primera

## Extreme small bowel resection in malrotation with acute midgut volvulus in a 3-year old boy: a case report

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### Izvleček

**Namen:** Malrotacija z volvulusom tankega črevesa je zelo redka prirojena anomalija, ki se z bruhanjem žolčne vsebine največkrat pokaže kot akutno stanje v neonatalnem obdobju in potrebuje takojšnje kirurško ukrepanje. Redkeje se simptomi pojavijo šele po prvem letu starosti, ko je zaradi neznčilne klinične slike težko postaviti pravilno diagnozo, zaradi česar lahko s pravočasnim zdravljenjem zamudimo. Kronični volvulus, ki je povezan z malrotacijo, se lahko zaradi torzije zgornjih mezenteričnih žil vsak trenutek poslabša, tako da nastane akutna venska kongestija in arterijska insuficienca. Samo takojšnja diagnoza in kirurško ukrepanje lahko preprečita odmrtje celotne stene prizadetega dela črevesa.  
**Poročilo o primeru:** Predstavljamo primer tri letnega dečka, ki je imel

### Abstract

**Purpose:** Malrotation with midgut volvulus is a rare congenital anomaly that generally presents acutely in the neonatal period, with bilious vomiting, and requires surgical intervention. It is rarely observed beyond the first year of life, when the diagnosis can be difficult because of intermittent symptoms and vague clinical findings, which can subsequently delay appropriate treatment. Chronic volvulus associated with malrotation has the potential to turn into acute vascular congestion and arterial insufficiency secondary to torsion of the superior mesenteric vessels. Early diagnosis and surgical treatment are essential to prevent progression to transmural bowel infarction.  
**Case report:** We present the case of a 3-year-old boy with a 1-year history

eno leto trajajoče težave v smislu ponavljajočih se bolečin v trebuhu in bruhanja, zaradi česar je bil fantek večkrat obravnavan v drugi zdravstveni ustanovi, vendar brez dokončne diagnoze. Nazadnje je bil deček z znaki akutnega abdomna in septičnega šoka nujno premeščen v našo ustanovo. Naredili smo urgentno laparotomijo in našli na volvulus zaradi malrotacije, z znatno nekrozo tankega črevesa, zaradi česar je bila potrebna obsežna resekcija prizadetega dela prebavne cevi. Po operaciji ni bilo zapletov in trenutno - eno leto po posegu - je deček na totalni parenteralni prehrani, raste primerno, uživa navadno hrano, drisk nima, prav tako ne znakov nezadostnega delovanja jeter. Čeprav je kvaliteta njegovega življenja nekoliko okrnjena, lahko skoraj nemoteno izvaja vsakodnevne aktivnosti in tako ne zaostaja za svojimi vrstniki.

**Zaključek:** V prispevku razpravljamo o pasteh pri diagnostiki malrotacije, ki so prisotne v primeru, če se simptomi tega redkega obolenja izjemoma pojavijo šele v otroštvu, o težavah pri zdravljenju, s katerimi smo soočeni ob razvoju akutnega abdomna in o terapevtskih možnostih pri bolniku po obsežni resekciji tankega črevesa.

of recurrent abdominal pain and intermittent vomiting, which was evaluated many times at other institutions without a definitive diagnosis. Finally, the boy was referred as an emergency, with signs of an acute abdomen and septic shock. Upon urgent laparotomy, midgut volvulus caused by malrotation with extensive intestinal necrosis was found, which necessitated an extreme resection of the small bowel. The postoperative course was uneventful, and presently, 1 year after the operation, the boy is on total parenteral nutrition, thrives normally, eats regular food, and has no diarrhea or signs of liver failure. Although there is some impairment in his quality of life, he can practice almost completely normal daily activities for a child of his age.

**Conclusion:** In this article, we discuss the diagnostic pitfalls that can occur when symptoms of midgut malrotation develop beyond the neonatal period, the difficulties of management in an emergency setting, and therapeutic options for the patient after massive small bowel resection.

## INTRODUCTION

Midgut malrotation (MM) is an anomaly of fetal intestinal rotation. The true incidence of rotational anomalies of the midgut is difficult to determine. Estimates in the literature range from 1 in 200 to 1 in 6000 of all live births (1). Differences in estimations suggest that many people with MM remain asymptomatic throughout life and their condition might never be diagnosed, or it is found coincidentally during diagnostic work up, laparotomy or autopsy for an unrelated disease (2). Symptomatic patients present either acutely with high intestinal obstruction due to midgut volvulus (MV) and/or Ladd's bands that partially obstruct the duodenum, or chronically, exhibiting a variety of less impressive, atypical and frequently nonspecific gastrointestinal symptoms (3, 4). MM is usually diagnosed in newborn and young infants; up to 75% of symptomatic cases occur in newborns, and up to 90% of symptomatic cases occur within the first year of life (5). The classic description of intestinal

malrotation is that of the term infant who presents with bilious emesis that prompts closer examination for duodenal obstruction. The upper gastrointestinal contrast series confirms the diagnosis by identifying the right-sided position of the duodenojejunal junction or evidence of MV. Unfortunately, diagnosis and treatment of intestinal malrotation might not always be so straightforward. In rare cases, congenital MM can be asymptomatic during infancy and show up with acute or recurrent intestinal symptoms later in childhood, adolescence or even in adulthood. Malrotation that presents beyond the neonatal period is associated with a multiplicity of symptoms, which often are nonspecific, and consequently, are associated with delays in diagnosis. MV can have devastating consequences at all ages, including the need for emergency surgery for smallbowel necrosis, extreme short-bowel syndrome with dependence on total parenteral nutrition (TPN), or even death (1 - 10).

Here, we describe the case of a 3-year old boy who had a 1-year history of intermittent abdominal pain and vomiting without definitive diagnosis and finally presented as an emergency with acute MV and massive small bowel necrosis caused by MM.

## Case report

A 3-year-old boy was referred to our institution late at night as an emergency from another hospital, with acute abdomen and septic shock.

The boy was born at term without any symptoms. He was healthy without any gastrointestinal disorder until he was 2 years old. He began complaining of recurrent abdominal pain accompanied by intermittent vomiting of ingested food, which was rarely followed by emesis of bile. After a day, the symptoms disappeared spontaneously, but they returned approximately once every 2 months. Over the 1-year period during which these -chronic- symptoms appeared, diarrhea sometimes occurred. The first presentation of the symptoms was at the time when the boy was traveling with his family to the far east. When they returned home, he was hospitalized several times; diagnostic work-up included allergy testing to foods, a lactose breath test, sweat chloride testing, stool guaiac, abdominal x-ray and ultrasound. All study findings were reported as normal. The boy thrived normally. The symptoms were interpreted as psychological disorders. One month before acute presentation, the symptoms appeared more frequently.

During the night, 24 hours before admission to our institution, the boy's symptoms, which were - well known - to his family, reappeared: colicky abdominal pain and vomiting. He had experienced these symptoms several times in the past, therefore, the family simply waited for them to cease spontaneously. This occurred faster than ever before (in approximately 6 hours). By morning, the boy felt no pain, although he was a little tired. He ate and passed normal stools. In the afternoon, the - well - known - symptoms returned, but now his abdomen distended as never before. He was admitted to another hospital.

Symptoms progressed rapidly, and vomiting of ingested food and bile continued, with hematemesis. An abdominal x-ray showed multiple airfluid levels in dilated small bowel loops (Fig. 1). He was referred to our institution. The boy looked severely ill; metabolic acidosis (pH - 7.2) with septic shock and severe anemia (hemoglobin: 69 g/L) were apparent. His abdomen was distended and painful, and there were no bowel sounds on auscultation.



**Fig. 1.** An abdominal x-ray showed multiple airfluid levels in dilated small bowel loops.

During emergency laparotomy MV due to MM with extensive intestinal necrosis was found (Fig. 2). The duodenum did not cross the midline and the duodenojejunal junction lay completely on the right side. The superior mesenteric vein (SMV) was to the left of the superior mesenteric artery (SMA) instead of to the right. The small bowel mesentery had a narrow base and it was thickened with lymphatic and venous congestion. There was also a thrombosis of the SMV (Fig. 3). The cecum was located at the lower part of the liver. All parts of the small bowel (as well as the fourth part of the duodenum and cecum) were obviously necrotic and therefore had to be resected. The coloduodenal bands were divided. A primary anastomosis was performed between the third part of the duodenum and the ascending colon. In addition to a nasogastric tube, a nasoduodenal tube and a colostomy tube were inserted to protect



**Fig. 2.** During emergency laparotomy, MV with extensive intestinal necrosis was found.



**Fig. 3.** Thrombosis of the SMV.

the anastomosis. The postoperative course was uneventful, and a contrast study revealed a patent anastomosis with no leaks.

Presently, 1 year after the operation, the boy is on TPN, thrives normally, eats regular food with no dietary restriction, passes pap-like stool once or twice daily, with no diarrhea, and has no signs of liver failure. Nine months after the operation, he developed catheter-related sepsis, which was treated successfully with catheter exchange and antibiotics. There is some impairment in his quality of life, but until now, he can be disconnected from TPN for 6 hours/day, and practice almost completely normal daily activities for a child of his age.

## DISCUSSION

Our report emphasizes the need to consider the diagnosis of MM in older children who have chronic abdominal symptoms. The difficulty of diagnosis lies in both the absence of specific physical findings and the low frequency after the neonatal period (1 - 10).

In the 20 years from 1985 to 2005, 34 neonates and children (including five neonates in whom MM was associated with other congenital anomalies) were operated on for malrotation in Slovenia, which means an average of 1.7 Ladd's procedures per year

(6). The proportion of children with MM who were  $\geq 1$  year old was only 20% in this series, which is within the range of 10 - 28% reported by others (3, 9). Malek has demonstrated how rare is symptomatic MM in children beyond the first year of life. In his multicenter analysis from an administrative database, which contained all pediatric discharges from 27 states in the United States, he found that children aged  $\geq 1$  year with malrotation were treated in only 4% of hospitals in the database, with most admitting only one child per year (5).

Our patient was asymptomatic until he was 2 years old. Presumably, when the symptoms of abdominal pain and vomiting of ingested food appeared for the first time, MV was present with intermittent torsion. The onset of symptoms and their severity depends on the degree and duration of vascular occlusion, which is characterized as "on and off" volvulization (3). In Prasil's study, the most frequent complaints in children older than 2 years were non-bilious vomiting and abdominal pain (7). A recent study by Penco and colleagues has demonstrated that the absence of classic presentation of MV with bilious vomiting and pain in children over the age of 2 years frequently leads to delay in the diagnosis of malrotation (8). Furthermore, disturbances in stooling pattern, which are present in 16 - 23% of reported cases, can be confusing, as in our case (9).

Unfortunately, the child developed symptoms for the first time when he was traveling abroad, thus the attention of the primary care physician at that time was primarily on food poisoning or allergy. In addition, after an acute attack, when he actually came to the physician, the child looked well, without complaints, and on physical examination, the abdomen was soft and non-tender to palpation. Symptoms in children are often mistaken for milk allergy, malabsorption, celiac syndrome or even psychological disorders (3). This happened in our case; in spite of several hospitalizations for possible gastrointestinal or psychiatric diseases, no definitive diagnosis had been made.

Abdominal ultrasound (US) was read as normal. The presence of inverted superior mesenteric vessels, a "whirlpool" sign, or duodenal dilatation on abdominal US have been shown to be sensitive diagnostic criteria for malrotation in children, but the examination is highly operator-dependent and is not secure enough to exclude the diagnosis (2). In a series of 107 patients operated on for malrotation as a primary condition, abdominal US was read as normal in 54% of cases (7). Given the concern with increased radiation exposure associated with computed tomography in pediatric patients, an upper gastrointestinal series, with or without barium enema, remains the preferred imaging modality for diagnosing malrotation in this population (2). The accuracy of the upper gastrointestinal series is reported to >80% (4). In a study by Prasil and colleagues, upper gastrointestinal series without barium enema was diagnostic in 94% of cases (7). In our case, the child was not sent for an upper gastrointestinal series.

With chronicity, there is a danger that a diagnostic label such as psychogenic can be attached to the patient, as physical findings are usually minimal, thus inhibiting intellectual vigor when - another attack - occurs (3). Long average delays of 1.7, 2.3 and up to 5 years in the diagnosis of malrotation in symptomatic children after infancy are suggestive of the challenges of diagnosing the condition in this population. Delay in diagnosis and surgical intervention remains the rule rather than the exception (8 - 10). It has been

reported that children beyond the neonatal period who present with MM frequently have potentially life-threatening complications of this anomaly; nearly half of symptomatic patients are admitted as an emergency, and the rate of volvulus with intestinal ischemia is 15 - 22% (3, 5, 7). When strangulation of the bowel developed in our case, the abdomen became distended and tender to palpation. Multiple airfluid levels in dilated bowel loops are ominous signs and usually indicative of established intestinal infarction (8).

During laparotomy, we exposed the twisted infarcted small bowel, which was obviously gangrenous. Thrombosis of the SMV probably worsened the situation and led to a shorter time interval of progression of ischemia to transmural necrosis. Although it is essential to preserve as much bowel length as possible, preferably including the ileocecal valve, we had no opportunity to preserve any length of the necrotic small bowel, except the first three parts of the duodenum. To have left any part of the necrotic small bowel in the abdomen, with the possibility of avoiding short bowel syndrome (SBS), would in our case, probably have led to further progression of septic shock and a consequent risk to the patient's life. In a study by Malek of seven patients aged >1 year, who were undergoing intestinal resection for acute MV, a 10-year old boy died on the day of admission after operation. Significant morbidity and mortality rates have been reported in other studies as well, when intestinal resection was needed in acute MV (3, 5 - 7). No data have been found in the literature, therefore Malek estimated a probability of 10-40% that a patient with MV would require intestinal resection and develop SBS that required TPN (5).

At present, TPN and small bowel transplantation remain the only viable therapeutic options for extreme SBS. However, sepsis and liver failure associated with TPN, and limited availability of donor organs and high graft rejection rates associated with transplantation, can limit their use (11). Attempts to engineer the small intestine have achieved varying degrees of success in animal models but at present are still experimental (12).

## CONCLUSION

In conclusion, we hope that enhanced awareness of the presentation of MM in older children will lead to shorter time to diagnosis and improved therapeutic outcomes in this rare, but potentially life-threatening disease.

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