



Intestinal malrotation and volvulus in adult life



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ABSTRACT

INTRODUCTION: Midgut volvulus due to intestinal malrotation is a rare cause of intestinal obstruction when occurring in adult life. This paper documents the difficulties in reaching an early diagnosis.

PRESENTATION OF CASE: We describe the case of an 85-year-old man with non-specific abdominal complaints for 20 years, who presented with sudden onset central abdominal pain. An acute median laparotomy under general anaesthesia was performed during which a 360° clockwise rotation of the small bowel around the mesenteric pedicle of the superior mesenteric artery and vein was found.

DISCUSSION: Malrotation is considered any deviation of the normal rotation of the midgut in embryological development, causing intermittent episodes of gastrointestinal obstruction or acute events of midgut volvulus. Although mainly a paediatric diagnosis, some cases do present in adult life. Radiologic investigations include: upper gastrointestinal contrast studies, Doppler sonography and a contrast enhanced CT of the abdomen. If a true malrotation is diagnosed or found by coincidence, a Ladd's procedure is advised, even if the patient is asymptomatic. There is no proven surgical strategy for preventing the recurrence of small bowel volvulus in case of malrotation of the midgut.

CONCLUSION: Non-specific recurrent abdominal complaints in adults of any age should raise suspicion of the possibility of a midgut malrotation or malrotation with or without intermittent volvulus. This case highlights the importance of diagnosis in an early stage.

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1. Case presentation

An 85-year old male presented with sudden onset central abdominal pain, progressive abdominal distension, diarrhoea, nausea and vomiting. Physical examination revealed a distended abdomen with tenderness in the upper quadrant and signs of circulatory shock.

The patient had been experiencing similar episodes of recurrent abdominal pain and vomiting for nearly 20 years and had therefore been under the surveillance of a gastroenterologist. Over the years, the patient underwent several diagnostic investigations: including upper gastrointestinal contrast studies and a computed tomography (CT) scan of the upper abdomen. However, no actual cause for the repeated episodes of abdominal pain was found. This led to a laparoscopic resection of the gallbladder in 2009.

An abdominal radiograph at presentation showed features of a distended small bowel. This was subsequently confirmed via an abdominal ultrasound and an abdominal computed tomography

(CT) scan without contrast. In retrospect a whirl sign was detected, suggesting a volvulus (Fig. 1).

A median laparotomy under general anaesthesia was performed during which a 360° clockwise rotation of the small bowel around the mesenteric pedicle of the superior mesenteric artery and vein was found (Fig. 2). The small bowel was oedematous and purple in colour (Fig. 3). The colon was positioned normally, with the caecum in the right lower quadrant. Some mesenteric adhesions were found. After counter clockwise derotation the bowel seemed viable after several minutes, so no bowel resection was performed.

The following day the patient did not improve and required inotropic medication. Lactate levels rose to 10 mmol/l, suggesting bowel ischaemia for which a second-look laparotomy was performed. This showed a viable bowel, although there were some bluish patches in the terminal ileum. No resection was performed.

After a short stable period, the patient's condition deteriorated rapidly on the 5th postoperative day. The intra-abdominal pressure rose to 26 mm Hg suggesting an abdominal compartment syndrome leading to severe abdominal sepsis and multiple organ failure. An urgent laparotomy was performed to relieve the intra-abdominal pressure. This showed an ischaemic terminal ileum of which about 90 cm (35.43 in.) was resected. The rest of the small bowel seemed vital enough. The abdomen was closed with a vicryl mesh.

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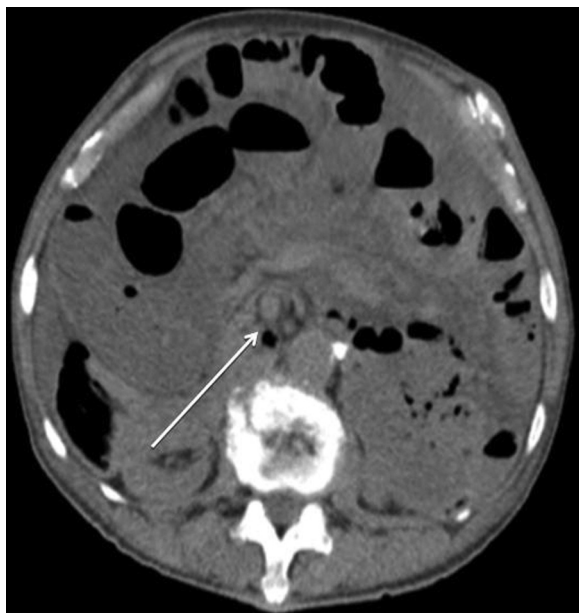


Fig. 1. An abdominal computed tomography (CT) scan without contrast showed a whirl sign (arrow), suggesting a volvulus.

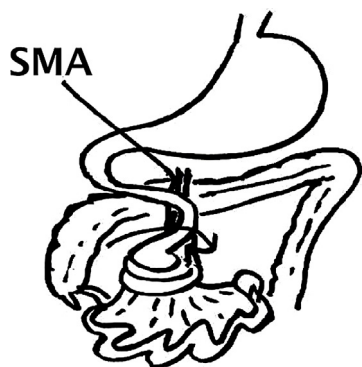


Fig. 2. Schematic reproduction of the 360° clockwise rotation of the small bowel around the mesenteric pedicle of the superior mesenteric artery (SMA).

Postoperatively, the patient was treated in the ICU with antibiotics for 8 days. Gastrointestinal bleeding due to gastritis and hospital-acquired pneumonia complicated his recovery. After 2 weeks the patient could tolerate enteral feeding completely. However, while still in hospital, the patient developed new similar non-specific abdominal complaints. A CT scan could not reveal any objective and/or obstructive cause for his complaints. Due to the patient's very poor physical condition, which was aggravated by



Fig. 3. The small bowel was oedematous and purple in color.

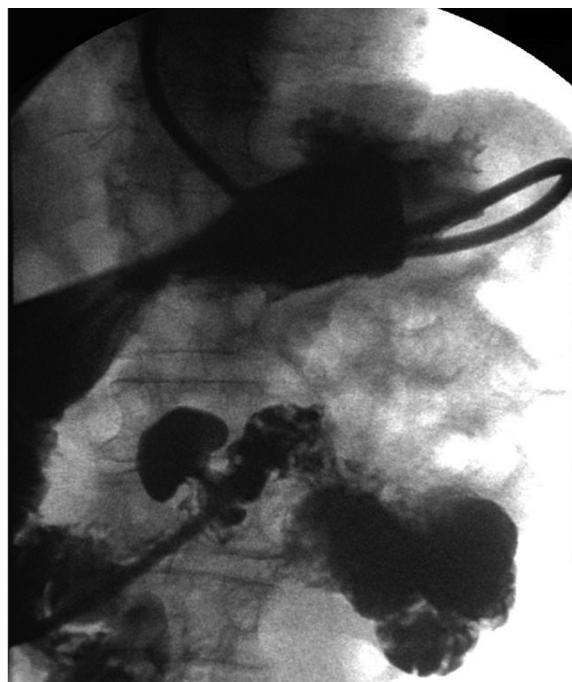


Fig. 4. Upper gastrointestinal contrast series showing a significant amount of jejunal diverticulosis.

his depressive state, he slowly deteriorated and died on the 48th postoperative day.

2. Discussion

Malrotation of the midgut has usually been estimated to occur in approximately one in 500 newborns¹ and presents within the first month of life in 64–80% of patients.^{1,2} However, some patients will present later, even in adulthood, or remain asymptomatic for life. Atypical symptoms such as sudden abdominal pain with (bilious) vomiting over a period of months or years are typical and may eventually lead to further diagnostics.^{3,4} Although several reports describe the presentation of malrotation in adult life, no cases are described occurring this late in life.

Intestinal malrotation is defined as any deviation from the normal 270° counterclockwise rotation of the midgut during embryologic development, also known as nonrotation.^{1,5} The failure of the normal physiological rotation of the midgut leads to various anomalies. Most commonly, the entire small bowel remains on the right side of the abdomen, while the caecum remains on the left, as a result of the absence of a ligament of Treitz. The caecum however, remains attached to the right side of the abdominal wall through persisting peritoneal fibrous bands (known as Ladd's bands). These bands often entrap the descending duodenum causing intermittent gastro-intestinal obstruction. Alternatively, the intestines can be situated correctly except for a small vertical attachment of the small bowel mesentery resulting in limited fixation to the retro peritoneum. This makes the small bowel highly mobile and therefore prone to midgut volvulus.^{1,5–8} This anomaly is known as a malfixation. During the operating procedure, no anomalies indicating a complete malrotation were seen in our patient. Therefore in retrospect, a malfixation was suspected to be the cause of this specific case of small bowel volvulus.

Diagnosing a malrotation or malfixation is exceptionally difficult. Upper gastrointestinal contrast series are the diagnostic method of choice in paediatrics.^{9,10} In our patient, a significant amount of jejunal diverticulosis was evident (Fig. 4), suggesting an obstruction located more distally in the gastro-intestinal tract. In cases of malrotation, a contrast enhanced CT scan shows an



Fig. 5. Abdominal CT-scan showing a SMA–SMV inversion with the vein (white arrow) rotating around the artery (blue arrow).

inversion of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV). In malrotation, the SMV is often situated to the left of the artery or rotates around the artery.^{11,12} In this case, on a CT scan on the 29th day after first presentation and operation, a clear SMA–SMV inversion was detected with the vein rotating around the artery (Fig. 5).

Midgut volvulus is easier to detect. Upper gastrointestinal contrast series show a typical corkscrew appearance of the proximal small bowel.^{3,5,10} However, in older patients with acute symptoms, a plain CT is generally performed in preference to contrast studies. The CT whirl or whirlpool sign describes the swirling appearance of bowel and mesentery twisted around the axis of the superior mesenteric artery.¹² Doppler sonography gives a similar image.¹³ Additional CT findings include duodenal obstruction, congestion of the mesenteric vasculature, and the evidence of underlying malrotation with a malpositioned duodenum and caecum.¹⁰ In our patient, beside the typical features of a distended small bowel, in retrospect a whirl sign was detected (Fig. 1).

There are no reliable means of identifying which group of patients with intestinal non/malrotation will develop subsequent complications of midgut volvulus.¹⁴ Many authors are therefore now advocating early surgical treatment for asymptomatic patients who are by coincidence diagnosed with any sort of malrotation.^{3,4,6,15,16} Intervention comes in the form of a surgical Ladd's procedure.⁸ This procedure consists of 4 parts: counterclockwise detorsion of the midgut volvulus if present; division of Ladd's bands overlying the duodenum thereby relieving the cause of the intermittent obstruction; widening of the narrowed root of the small bowel mesentery by mobilizing the duodenum; and division of the adhesions around the SMA to prevent further volvulus.^{5,8} In case of a malfixation and a normally located small bowel, no means are identified to surgically prevent midgut volvulus. There is a high risk of these patients developing complications in the future. Some authors state that the use of Ladd's procedure or division of Ladd's bands and adhesiolysis relieves symptoms and due to the formation of intra-abdominal adhesions after the procedure, prevents recurrence in the majority of patients.⁵ However, in the case of our patient, there was just the narrow vascular pedicle for which no preventive surgical treatment is documented in the literature.

Conflict of interest

None declared.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and case series and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors' contributions

Bastiaan W. Haak (corresponding author) assisted in the surgical procedures; conceived the write-up; performed the literature search and concluded the manuscript preparation. Sander T. Bode-witz: responsible for interpretation of the radiographic images; participated in the write-up concerning the diagnostic approach to malrotation and volvulus. Caroline F. Kuijper participated in the write-up concerning the surgical approach to malrotation and volvulus. Louise M. de Widt-Levert: treating physician; performed the surgical procedures; involved in the pre-operative work-up and post-operative care of patient; participated in the write-up; assisted in the manuscript preparation.

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