

Complicated Adult Midgut Malrotation: A Challenge to Diagnose and Treat

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1. Abstract

Midgut malrotation (MGM) refers to a broad spectrum of rotation and fixation anomalies of the gut during fetal development, which are clinically evident mostly early in life as newborn emergencies and rarely later as milder forms in adults. Most commonly, adult patients are asymptomatic throughout life or they are insidiously presenting unspecific symptoms for a significant period of time before the diagnosis is made. Less commonly, adult MGM cases are acutely complicated with challenging and life-threatening conditions, such as the obstructive midgut or right colon volvulus and the ileocolonic intussusception, which require prompt diagnosis, intensive resuscitation and emergent surgical intervention. Intentionally, high index of suspicion and awareness are essential, if bowel infarction is to be prevented and high morbidity and mortality reduced.

Keywords: Adult midgut malrotation, midgut volvulus, right colon volvulus, ileocolonic intussusception, bowel obstruction, diagnosis, surgical treatment

2. Introduction

Midgut malrotation (MGM) refers to a wide spectrum of congenital abnormalities occurring as a result of partial or complete failure or arrest of the normal 270° intestinal rotation and fixation process during embryonic period, which may present within the peritoneal cavity as abnormal position of the intestines, especially the right colon, the small intestine with its mesentery, and even the duodenum¹⁻⁴. This congenital abnormality is usually an isolated condition with an incidence in general population one for every 200-500 newborns and symptomatic cases occurring in one to 6000 newborns, sometimes it is associated with congenital heart disease or situs or biliary tract anomalies; it is mostly recognized and treated by pediatric surgeons^{1,2,5}. In adulthood, MGM is estimated to occur in approximately 0.2%-0.5%^{2,5,6}, and is clinically significant in only a small portion of cases. The rate of

incidence is approximately the same for men and women⁷. Indeed, most adults are asymptomatic and incidentally diagnosed later in life at surgery for other diseases. Some patients may present chronically with nonspecific symptoms (i.e. vague abdominal pain) or recurrent intermittent bowel obstruction, and a minority with acute or fulminating bowel obstruction due to caecocolic mobility and torsion or even bowel invagination, which may lead to bowel necrosis^{3,8-11}. Adult caecocolic torsion is a class of right colon volvulus (RCV), and MGM is always associated with an increased risk of this devastating complication^{4,7,9}. The more rare bowel invagination in cases with underlying MGM, namely ileocolonic intussusception (ICI), may present independently or coexisting with RCV^{3,9,11}.

The wide range of rotation and fixation defects ascribed, resulting in accordance to the stage at which the embryologic error occurred, is the explanation of the wide and more obscure constellation of clinical symptoms during adulthood, and the reason why the diagnosis of MGM is often delayed^{4,12}. In addition, the index of suspicion for MGM progressively decreases in the older population and, thus, the clinical diagnosis is not considered in the initial evaluation^{1,12}. This article seeks to highlight the importance of a thorough understanding of the undiagnosed rotation and fixation midgut aberrations in the adult abdomen in cases when these abnormalities are complicated and particularly presented with the features of a surgical emergency.

3. Discussion

3.1. Pathophysiology

During the 4th to 10th weeks of intrauterine normal life, the rapidly growing primitive midgut of the fetus herniates through the site of the future umbilicus (stage I). The midgut migrates back into the peritoneal cavity during the weeks 10th to 12th, with the caecocolic and duodenojejunal loops rotating 270° in a counterclockwise manner with respect to the superior mesenteric vessels; the duodenojejunal junction is anchored at the Treitz ligament behind and to the left of the aforementioned vessels, and the caecum is attached to the right iliac fossa (stage II). The final stage consists of fusion of various parts of the mesentery with the posterior parietal peritoneum (stage III)^{1,2,4,5,7}.

Most severe forms of MGM occur as a product of alterations in the normal embryonic gut development between 10-12 weeks of gestation⁵. MGM has been classified into three major types, according to the stage at which the embryologic error occurs. Type I is defined as non-rotation of colon and duodenum, and it is characterized by transposition of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV). Type II includes non-rotation or reversed rotation (paraduodenal hernia) of duodenum, and reversed rotation of duodenum and colon; it can manifest as duodenal or transverse colon obstruction. Type III includes non-rotation of colon - clinically presented as RCV- and duodenum,

incomplete fixation of the hepatic flexure or caecum (may manifest duodenal obstruction by Ladd's bands or caecal volvulus), and internal hernia near the Treitz ligament^{1,13}. Importantly, the complexity of the embryologic development of the gastrointestinal tract and mesentery provides a platform for the creation of variant abnormal membranes, veils and folds, collectively termed peritoneal or parietocolic bands (i.e. Ladd's bands, funicular Jackson's membranes), located in the regions of the duodenum, duodenojejunal or ileocaecal junction, the ascending colon, and the right hepatic lobe; subject to specific conditions, these deviant structures may act as a fulcrum of intestinal blockage or torsion⁹. Adult MGM, accompanied with prerequisites for bowel torsion, mainly the mobile redundant caecocolon-improper development of fusion of the mesentery of right colon with the parietal peritoneum-and the narrow based small bowel mesentery, can be complicated when the caecocolon along with terminal ileum and its mesentery twists, usually clockwise, around the SMA axis^{4,5,8,13}. This condition is termed RCV. Indeed, any transverse kink or torsion to the right colon will take time to impede the profuse anastomoses of the blood supply. The complete and persisting torsion results in mechanical lumen obstruction, blood supply compromise or cut-off, and finally frank intestinal necrosis and perforation^{4,5,7,12}. This is the most feared complication of MGM and a clear indication for emergent surgery. Even more rarely, another acute condition, namely ileocaecal or ileocaecal-colonic intussusception (ICI) in various degrees, may coexist with adult MGM. ICI is facilitated by the caecocolic mobility and the altered peristaltic bowel motion, the increase value in intraluminal pressure, and even the presence of a demonstrable ileocolic structural lesion(identified in 90% of cases), that serves as a "lead point" for (asymmetrical) invagination and it is neoplastic in up to 75% of cases, mostly malignant (synchronous "slide and twist" process)^{3,4,9,11}. Similarly, ICI can result in obstruction and subsequent bowel ischaemia⁹. Some hypotheses explaining the coexistence of ICI and RCV in rare adult cases are found in the relative literature^{3,11}, but possibly the exact mechanism is not quite understood.

3.2. Presentation and diagnosis

Whilst MGM is a well described cause of neonatal bowel obstruction and approximately 85% of these cases are presenting in the first two weeks of life, it is being increasingly accepted that its unrecognized persistence into adult life can cause diagnostic difficulty and uncertainty¹².

In adolescents and adults, milder forms of MGM than in infancy and childhood, and highly variable, can present and a prevalence of 0.2%-0.5% has been reported^{2,5,6}. In a review of the literature there were only 40 MGM cases described between the years 1923 and 1992². On the other hand, although RCV and ICI of any aetiology are uncommon causes of abdominal pain and bloating, as well as of bowel obstruction in adults, each one accounting for less than 1% of cases with acute bowel obstruction in western countries, they can both be challenging and life-threatening conditions, if not timely diagnosed and treated^{14,7,8}.

In general, most adults affected by MGM are asymptomatic, and their diagnosis may be obscure^{2,5,12,14}. The diagnosis

remains also difficult and challenging in some symptomatic patients, who frequently visit different hospitals, mostly with recurrent non-specific mild complaints, such as abdominal pain or constipation, or chronic diarrhea and even malabsorption, caused by chronic intermittent volvulus and chronic venous and lymphatic obstruction^{5,7,10}. Under specific circumstances, a minority of patients, having midgut malformations with indolent manifestation, may present with progressed acute or fulminating obstructive volvulus, that is usually a case of colonic and intestinal ischaemia running the risk of perforation or rupture^{7,8,10}. Predisposing factors have accordingly been implicated, such as intestinal atony, chronic intestinal pseudoobstruction, paraplegia with impaired colonic mobility, Hirschprung's disease, chronic constipation, laxative abuse, pregnancy and distal obstructing tumour^{1,3,4,8,13}. On the other hand, ICIs may rarely coexist with RCV and MGM, but they are usually malignant and consequently have clinically important differences from their small bowel counterparts^{3,9}. Both acute RCV and ICI have similar clinical symptoms of bowel blockage, but the symptomatology depends on the type of variant produced by the torsion or the invagination respectively.

For chronically symptomatic, uncomplicated MGM a diagnosis can be made with upper gastrointestinal contrast enhanced follow-through study, which may show the typical "corkscrew appearance" caused by the dilatation of various duodenal segments and the relocation of the duodenal junction and the jejunal folding^{1,5,7}. In cases complicated with recurrent RCV, ultrasonography/Doppler may show some degree of rotation of SMV around SMA ("Whirl or Whirlpool sign") and an abnormal location to the right of the jejunal arteries; angiography may show similar findings^{5,7}. In chronic cases, computed tomography (CT) scan and magnetic resonance imaging can reveal abnormal position of the right colon to the left and of the small bowel loops to the right, changes in duodenal anatomy and relationships, and some swirling appearance of bowel and mesentery twisted around the SMA axis("Whirlpool sign"),evident of underlying MGM^{1,4,5,7-9,12,14}. Notably, sectional imaging studies are incapable of identifying adhesions among bowels. In acutely presented MGM cases complicated with RCV, similar and more intense and revealing findings on radiological evaluation and CT scan can be seen. Radiographies may show a large gas-filled ahaustral colon extending from the right pelvis to the left upper abdomen ("coffee bean deformity") and, contrast-enhanced, the site of torsion ("bird's beak sign", "column cut-off sign")^{9,13}. CT scan in the emergency service has proven to be effective and highly accurate (up to 90%) for RCV, sometimes providing additional anatomic clarification, which may alter the management plan (Fig. 1). In acute RCV, CT scan may demonstrate loops of either distended or collapsed bowel and its mesentery sequestered to the right, characterized by congested mesenteric vasculature and twisted around their vascular pedicle, and can identify the location of a closed loop obstruction -helpful in discriminating right colon from sigmoid colon volvulus- or concomitant pathology that may have lead to the onset of volvulus^{1,3-5,7,9,12,14}. An ominous sign should be the presence of intestinal ischaemia or necrosis. However, the presentation of adult MGM can be obscure, even though the CT

scan “Whirlpool sign” may give suspicion of bowel twisting⁷. Finally, CT scan usually demonstrates the hallmarks “bowel-within-bowel” and “target lesion” of ICI (Fig.1), which however is less specific if there has been ischaemia and perforation^{3,11}.

Delayed diagnosis of MGM is common in several case reports describing atypical presentations of left-sided acute appendicitis¹². Higher mortality rates are seen in cases with acute onset of midgut volvulus, delayed diagnosis, or the presence of bowel necrosis⁵. While a CT scan can remove much of the diagnostic uncertainty, the identification of MGM can be missed unless there is a high index of suspicion¹². It is well emphasized that, in acute presentations of MGM in adult patients, awareness of the clinician and the need for an early investigation and diagnosis, suitable preparation, and prompt expedient surgical intervention, are essential, if bowel infarction is to be prevented and high morbidity and mortality reduced.

3.3. Treatment

Techniques for non-operative relief of the bowel obstruction in complicated MGM cases who have not an absolute indication for urgent surgical intervention, which are absolutely discouraged in the setting of potential gangrene changes, should not be considered as definite therapy but can sometimes convert an emergent procedure in a poorly prepared patient to a semi-elective one, providing temporary mechanical detorsion and decompression of the bowel^{9,13}. However, in contrast to sigmoid volvulus, colonoscopic devolvulation to relieve RCV is rarely accomplished and revolvulation often occurs, and also leads to a high risk of perforation^{8,9}. It has been postulated that the potential to develop sudden onset of acute MGM complications in asymptomatic patients, at any age, always exists. Intentionally, several authors advocate a surgical correction in all patients with a diagnosis of MGM, regardless of the presence of symptoms and regardless of patient's age^{7,14}.

The preoperative evaluation of adult patients with acute abdomen due to complicated MGM, who are often elderly and debilitated, should include an assessment of their current physiologic disturbance and bowel function and an anal sphincter examination; this former practice may influence intraoperative decision making, as the patient with poor rectal tone and incontinent sphincter control might be better helped with an ostomy as opposed to anastomosis after bowel excision⁹. What is primarily required in management is the immediate and intensive fluid resuscitation and hemodynamic support for the unstable patients and the variable supporting for the remaining, coupled with a simultaneous administration of broad-spectrum antibiotics (potential bowel ischaemic necrosis? need for bowel excision?), and laboratory work-up^{4,5}. Emergent exploratory surgical intervention should never be delayed in unstable patients in the fraudulent hope of correcting firstly hemodynamic instability, because its reversal will not occur until complete bowel obstruction and strangulation have been timely resolved (Fig.2).

The classic treatment for the neonatal MGM, the Ladd's procedure, entails division of the abnormal adhesive coloduodenal bands tethering the midgut and causing duodenal/bowel compression,

untwisting in a counterclockwise direction of any MGM volvulus (if present), division of the inter-mesenteric bands, section of adhesions near the superior mesenteric vessels in order to limit their rotation and broaden the mesenteric base, setting segments of colon and small bowel to neutral position, and removal of the malpositioned vermiform appendix^{2,5,7}. Ladd's procedure can also electively be performed by laparoscopy^{2,7}; Mazzioti et al⁶ described a method to assure the possibility of laparoscopic Ladd's procedure, on every occasion that the length between the duodenojejunal junction and the ileocaecal valve is less than half the transverse diameter of the peritoneal cavity. However, in the emergency setting in adolescents and adults, the single Ladd's procedure, being the standard interventional repair for early in life and elective cases, is usually not a surgical option since bowel resection is mandatory. In the case of acutely presented adult MGM, which is more commonly complicated with complete RCV associated with bowel ischaemia or necrosis and that may require extensive bowel resection, a Ladd's procedure type adhesiolysis and the setting of the bowel to neutral position may complete the basic operation^{4,5,7}. Laparoscopic approach for adult acute RCV (or ICI) with complete bowel obstruction has very rarely been reported, and this is probably due to the rarity of the condition and perceived difficulties with using laparoscopic tools^{2,13}.

In general, acute complicated adult MGM requires emergent surgery, and not a single surgical option can be used, that is critical to tailor the approach to the individual patient's status for surgery and the type and stage of the presenting variant^{8,9,13}. Recommendations for operative approaches in emergent surgery are:

On stable patients, who are adequately resuscitated, with no bowel ischaemic necrosis or perforation or presence of highrisk factors, the optimal surgical management entails open right hemicolectomy with primary ileotransverse anastomosis for RCV, and en block oncologic resection, without manipulation for reduction, and anastomosis for the rare (coexisting) ICI. Non-resectional approaches in the setting of viable colon after devolvulation, such as the colopexy, have been reported to be associated with a recurrence rate as high as 40% and a mortality up to 18%^{8,9,13}. On the other hand, the occurrence of a lesion-“lead point” for ICI, which is more likely malignant (i.e.caecal adenocarcinoma), sets out not only the type of bowel excision but also alters the line of the subsequent management^{3,9,11}. On debilitated- malnourished elderly patients with severe comorbidities, who require either urgent or semi-elective surgery but they are at extremely high risk for bowel resection or anastomosis, alternatives that have been proposed as suboptimal surgical option but live saving measures in cases without gangrene include mainly variations of (colo) pexy/fixation and ostomy (i.e. caecostomy)^{9,13}.

On unstable patients with sepsis from bowel ischaemia or perforation or complete refractory obstruction (strangulation), who have no time for full resuscitation and optimization, treatment principles include immediate transfer to the operating room under simultaneous resuscitation and surgical resolving of the primary bowel obstruction, removal of the ischaemic bowel with control

of spillage, and proximal bowel diversion (usual)^{4,8,10,13}. For the emergent patient *in extremis*, characterized by one or a combination of metabolic acidosis, hypoxia, sustained hypotension and coagulopathy, “damage control” surgery includes rapid stapled in situ resection of the involved pathologic bowel segments (either volvulised gangrenous, or intussuscepted with oncologic requirements) and their mesentery (right hemicolectomy), debridement/lavage to remove gross contamination, enterostomy (instead of anastomosis), and temporary abdominal closure⁹. In these cases, based on the degree of postoperative response to resuscitation, a return to the operating room within 24-48 hours for possible bowel reconstruction or further “damage control” closure (for bowel of questionable viability left in situ) or “second look” is needed. For the emergent patient who is *not in extremis* and responds to resuscitation a similar excision (i.e. right hemicolectomy) and peritoneal lavage for complicated RCV and/or ICI and a primary ileotransverse anastomosis can be performed, with the prerequisites of achieving viable and healthy bowel ends and no adverse factors of anastomotic healing (i.e. malnutrition, steroid use, etc.) exist⁹. In some cases, resection and end ostomy or even anastomosis (on apparently healthy bowel ends) and proximal (protective) loop ostomy are viable options.

An acute presentation of adult complicated MGM may sometimes be associated with excessive bowel necrosis and may lead to massive bowel resection, either during the index surgery or during a second look laparotomy for a non-recovered borderline ischaemic segment⁷. If survived, patients with short bowel syndrome require closed continuous treatment caring for malnutrition, electrolyte disturbances, immune deficiency, and even sepsis.

5. Figures

Figure 1. Emergent CT scan (axial images) in a 50-year old man (left) and an 88-year old woman (right) with acute abdomen -complete bowel obstruction, indicating, left: close-loop colonic obstruction and distended small bowel loops due to twisting of mobile (excessive) right colon/mesentery with vascular strangulation (arrow for “Whirlpool sign”); right: ileocaecal mass and displaced / twisted right colon, with distended bowel loops and gas-less left colon (black arrows for intussuscepted parts, white arrow for “Whirlpool sign”). These patients from our own material ^{3,4} underwent urgent resective surgery for: complicated underlying MGM with RCV (left) and RCV/ malignant ICI (right) (see, left and medium images in Fig.2)

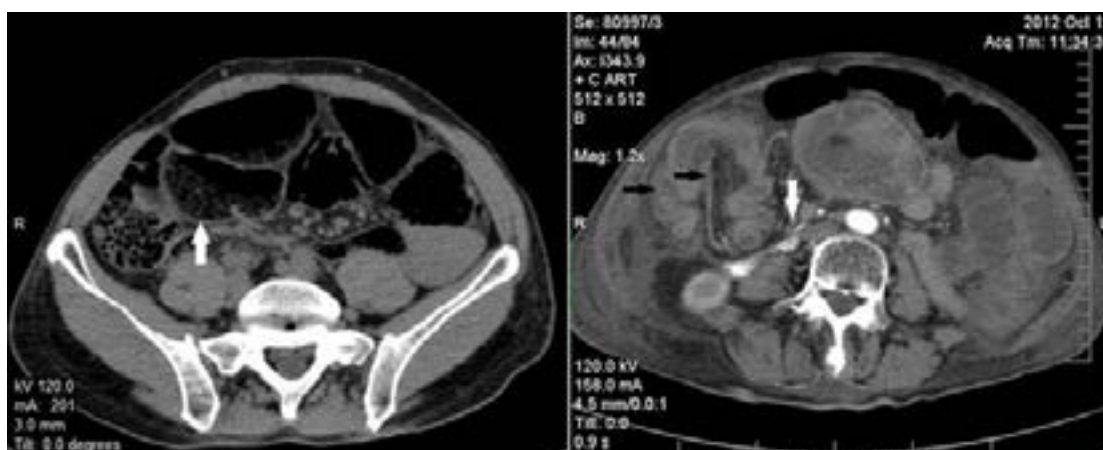


Figure 1

Figure 2. Intraoperative findings of complicated MGM, left: volvulised unfixed right colon (white arrowhead) with adhesive band between mesenteries (white arrow) and excessive dolichosigmoid loop (black arrowhead); bands connecting transverse colon/mesocolon to right hepatic lobe beneath are not shown; medium: malignant ICI with regional necrosis/inflammation (arrows) and de-volvulised unfixed right colon, covered with funicular membranes (arrowhead); right: unfixed volvulised right colon in a woman 46-year old (arrow shows v. appendix).



Figure 2

6. Informed consent statement

Written informed consents were obtained from the patients for presentation in Congresses or publications with use of images.

7. Conflict of interest

The authors declare no conflict of interest.

Institutional review board statement

This paper has been approved by the Scientific Committee of the Hospital in place of review board.

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