



# Complicated Midgut Malrotation in Different Ages: Diagnostic and Treatment Challenges. A Review

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## Abstract

Midgut malrotation (MGM) is a congenital abnormality occurring as a result of failure of the normal midgut rotation and/or fixation process during the embryologic development. The consequences and life-threatening complications of MGM, such as the midgut volvulus and duodenal compression, are familiar to Pediatric surgeons, but are rarely encountered by those caring for adults. Acute presentations require immediate resuscitation and emergent surgery to relieve bowel obstruction and prevent bowel necrosis. In most cases without bowel necrosis, a Ladd's procedure is the surgical intervention of choice. In emergent older cases, bowel resection is usually required. The prognosis of affected patients depends on the disease stage, comorbidities, timely diagnosis, and the type of surgery. Issues of concern with MGM is the ensuring of early diagnosis, the management of insidious or asymptomatic cases, and, on surgical cases, the extent of bowel resection, the construction of anastomosis, the prevention of recurrence, and the technique of abdominal closure. Adult presentations may pose diagnostic challenges, and clinical awareness is always required. This article reviews relative literature and seeks to highlight the importance of the thorough understanding of the presentation of complicated MGM throughout life, and make recommendations for its management.

**Keywords:** Midgut malrotation; Right colon volvulus; Bowel obstruction; Diagnosis; Surgery

## Introduction

Midgut (or intestinal) malrotation (MGM) is a congenital anomaly that occurs secondarily to the partial or complete failure of the midgut (MG) to rotate 270° counter-clockwise around the axis of the superior mesenteric artery (SMA) and to achieve retroperitoneal fixation during the embryonic development, and may present as abnormal position and altered mobility of the large and small bowel within the peritoneal cavity [1-7]. MGM encompasses a wide spectrum of clinical entities reflecting the various anatomic configurations of the MG and its consequences [4,5]. This benign clinicopathological syndrome affects predominantly the newborn and pediatric population, but adult cases have also been occasionally reported [4,8-11]. The true prevalence of MGM in all ages is difficult to define due to the unclear number of asymptomatic cases; a rate of 0.2% -1% of the population has been reported [5,12]. Furthermore, the incidence

will likely continue to increase as incidental diagnosis of MGM is increasing grace to advances in imaging [13]. It is 1% in autopsy studies [14], and an incidence of 1 in 500 live births has been reported, while symptomatic MGM occurs in 1 in 6000 live births [4,5,7]. It is classically thought that, around 90% of patients with MGM present in the first year of life, with the majority (64%-80%) presenting in the neonatal period or within the first month of life [5,12-15]. Beyond the neonatal period, MGM occurs with decreasing frequency [5,16]. Adult MGM presents similar trends as the older child, and it is usually an accidental radiological finding or, more rarely, occurs as an acute event, accounting for only 0.2%-0.5% of cases [11,13]. A 1.7:1 ratio of adult male to female preponderance has been reported [13]. Adult presentations may lead to diagnostic challenges and, if diagnosis of complicated cases is delayed, to worse outcomes [5,8,16-19].

Given the rarity of the MGM condition in older patients, there are still many barriers in the timely diagnosis and management,

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including the various vague clinical presentations, the provider's inexperience, and the uncertainty with the ideal treatment, all contributing to the increased morbidity and mortality observed with MGM in adults when compared to the pediatric population [5,8,20]. This highlights the need for both radiologists and clinicians to maintain a high index of suspicion for MGM if abdominal intestinal abnormalities happen to be identified incidentally, i.e., on a computed tomography (CT) scan, and to appreciate the challenges associated with this condition in the adult population, in order to ensure early management [5,13,16]. MG volvulus refers to the twisting of small bowel loops and the proximal colon around the SMA and their corresponding mesentery [6,7,17-19,21-23]. This acute caecocolic torsion is a type of right colon volvulus (RCV) [18]. It is the most fearful complication of MGM since it causes acute bowel obstruction, potentially leading to bowel blood compromise and ischaemic infarction/necrosis [5,13,14,18,19,21,23]. In MGM cases, MG volvulus is often ileocolic, accounting for 25% of all intestinal/colonic volvulus (10% in case of caecal volvulus), and it is as rare as 1-3% of all cases of bowel obstruction in pregnancy [16,23]. There are numerous reports of neonatal cases of MG volvulus in the literature, while intrauterine occurrences have been reported less frequently [6-8]. Instead, MG volvulus is far more common among the pediatric than in the adult population; only an estimated 15% of adult MGM cases present with MG volvulus [13,14,19,23].

This article seeks to highlight the importance of understanding the variable presentations of the MGM throughout life and to make recommendations for its management. We reviewed the literature for MGM and MG volvulus, and provided image material from cases of our experience. A thorough search via the PubMed and MEDLINE® databases has been performed using the keywords "midgut malrotation", "intestinal malrotation", "midgut volvulus", "complicated", and "emergent" with time limit mainly the past decade.

## Discussion

### Etiology - pathophysiology

The normal MG development is a complex process that requires intraabdominal 270° counter-clockwise rotation of the gut tube around the SMA axis during the 4<sup>th</sup>-10<sup>th</sup> /12<sup>th</sup> week of gestation [5,9,11,13]. Normally, the wide base of the intestinal mesentery, extending from the Treitz ligament to the ileocaecal valve, along with retroperitoneal attachments, prevent bowel twisting [6,7]. Rarely, MG aberrations are associated with congenital heart disease or visceral situs (inversus, ambiguous) or biliary anomalies [11,24].

The degree the bowel has failed to undergo its physiological rotation and fixation can vary widely, explicating the multiple

forms of MGM (Dott 1923); the most common are incomplete rotation, non-rotation and reversed rotation, with subtypes based on the orientation of the duodenum and colon [4,9,13,15,]. The non-rotation type on the one extreme of the spectrum is characterized by positioning of the small bowel in the right hemiabdomen and the large bowel in the left, absence of the Treitz ligament, malfixation of the narrow-based mesentery, inversion of the superior mesenteric (SM) vessels, and hypoplasia of the pancreatic uncinata process [5,13,15,]. On the other end, the most rare reversed rotation form results from the clockwise rotation of the MG in relation to the SMA, which leads to malposition of the transverse colon posterior to the SMA, prone to extrinsic obstruction, and of the duodenum anterior to the SMA [5]. Among the remaining MGM cases, constituting the majority, the incomplete rotation type is the classical MGM type that is usually described, and is due to bowel which rotates 180° counter-clockwise instead of 270°; this leaves the caecum in the right upper quadrant with a narrow vascular pedicle at risk of MG volvulus [4,5,15]. In another classification, MGM is divided into three major types: non-rotation (type I), duodenal malrotation (type II), and combined duodenal and right colon malrotation (type III) [24,25].

Accordingly, abnormal fibrous peritoneal bands (William Ladd, 1932) may firmly attach the right lateral abdominal wall and the liver to the caecum and/or ascending colon, and exercise external compression to the duodenum and jejunum, predisposing also to MG volvulus [4,5,13,15]. Obstructing Ladd's bands and congenital internal hernias (i.e., right paraduodenal hernia) may cause acute presentation in 10% of cases [5,12]. Other rare parietocolic or peritoneal membranes, veils and folds (i.e., funicular Jackson's membranes) may also act as a fulcrum for intestinal blockage or torsion [1,18].

The exact etiology of MGM remains uncertain. Some studies have suggested a genetic component, such as the association between mutations in the forkhead box transcription factor (FOXF 1) and L-R asymmetry genes [20]. MGM, as well as other gastrointestinal (GI) changes, have also been linked to chromosomal abnormalities, such as the trisomy of the long arm of chromosome 16 and a ring chromosome 4 [13].

The MG volvulus on underlying MGM occurs when the unfixed hypermobile caecum/ascending colon, along with the small bowel (ileum), are additionally and concurrently twisted greater than 180° around their own long and narrow mesentery and the SM vessels [1,6,7,13,16-19,21-23,26]. Based on autopsy and cadaveric studies, the fixation of the ascending mesocolon to the right gutter is absent in 10-15% of the population, a rate that far exceeds the incidence of acute RCV [21,22]. In practice, continuous bowel twisting may lead to extrinsic occlusion of the vasculature within the bowel wall, resulting in the ischaemic infarction/necrosis of the involved segment supplied by the SMA

and the poor venous return, and eventual bowel obstruction (closed loop obstruction), bowel perforation and peritonitis [3-6,13,15,19,23]. In adults, various predisposing factors for the initiation of this complication have been implicated, such as the chronic intestinal distension or atony, chronic constipation, paraplegia with impaired colonic motility, bowel hypoganglionosis or chronic intestinal pseudoobstruction, consequences from laxative abuse or high fiber diet, pregnancy, and distal obstructing tumour [9,17,22].

Another condition which is very rarely associated with MGM, presenting independently or coexisting with MG volvulus, is the ileocaecal /ileocolonic intussusception (ICI), which can also result in obstruction and bowel ischaemia [17,18]. ICI is facilitated by the caecocolic mobility, and, in adults, by the altered peristaltic bowel motion, the increased intraluminal pressure, and even the presence of a structural ileocolic “lead point” for invagination, a malignant mass in most of the cases [17,18].

### Clinical Presentation

The clinical image of MGM correlates with the age of presentation, with infants commonly presenting with acute MG volvulus, while this in contrast only accounts for 15-20% of adult cases [5,13]. Although acute obstruction secondary to MG volvulus or obstructing congenital bands can occur in all age groups, there is a tendency towards more chronic subtle or obscure symptoms as a child grows old [5,16].

The current literature suggests that around 90% of MGM cases present in the first year of life, the majority with acute bilious vomiting secondary to MG volvulus [3,5]. In neonates, the cardinal sign of bilious emesis facilitates the establishment of the diagnosis within hours to days; this is a surgical emergency for the neonate, as well as for older pediatric patients [3,4].

Bowel obstruction is rare in pregnancy with an estimated incidence of 1 in 10000; the main underlying etiologies include adhesions (60%) and volvulus (25%) [23]. In the prenatal-intrauterine period, maternal and fetal outcomes following maternal MG volvulus can be disastrous, especially if the diagnosis is delayed [6,23]. In their systematic review, Chong et al [23] demonstrated that, maternal and fetal mortalities were 13% and 35% respectively; all mother deaths happened during the third trimester. Since fetal outcomes are directly linked to the maternal physiology, delayed diagnosis of maternal MG volvulus and delayed surgery may lead to bowel infarction with associated hypovolaemia, renal failure and septic shock that result in fetal compromise [23]. MG volvulus in pregnancy is a surgical emergency, though commonly masquerades as symptoms of pregnancy, which may lead to delays in proper diagnosis and definite optimal treatment [7]. The classic triad of maternal MG volvulus consists of generalized abdominal pain, vomiting and constipation [5]. Similarly, in the very rare case of intrauterine

fetal MG volvulus [i.e., polyhydramnios in ultrasonography (U/S), “intraabdominal mass” in magnetic resonance imaging (MRI)], timely diagnosis and two emergent surgeries are required (see below) [7].

Adult presentations of MGM can be broadly split into incidental, chronic, and a minority of acute cases [5,10,13-15]. MGM is often found incidentally on asymptomatic patients who undergo imaging or laparotomy for other health conditions [13]. The chronic presentation of MGM tends to be insidious, with the patient experiencing nonspecific vague or subacute GI symptoms over a period of months to years [4,13]. In this case, common symptoms include crampy abdominal pain, bloating, nausea/vomiting, and weight loss (food intolerance) [13,14]. Patients may possibly experience intermittent or recurrent self-limited bowel obstruction episodes, attributed to either Ladd’s bands compression or MG volvulus [5,10,13,14]. Often, the diagnosis is missed for a considerable time and it is only achieved via CT scan or exploratory laparotomy [4,5,10,13,14]. Acute or acute on chronic presentations are less common, comprise only up to 20% of all adult cases, and are usually secondary to MG volvulus [5]. The common symptoms and signs of adult MGM presenting with MG volvulus are severe epigastric or umbilical pain, nausea/vomiting and abdominal distension, which may lead to bowel ischaemia and eventual bowel necrosis with potentially fatal consequences [5,13,14].

Even more rarely, MGM is complicated with ICI, which is a similar obstructing bowel condition [9,10,17,18]. Colonic carcinoma in the setting of (adult) MGM has been sparingly reported worldwide, mostly from Japan; among 59 reported cases, in 38 patients (64.4%) the tumour was located in the right colon [15].

### Diagnostic Evaluation

Timely intrauterine diagnosis of fetal MG volvulus may pose a challenge due to the absence of specific symptoms and signs. It is pertinent for radiologists, sonologists and obstetricians to be aware of fetal MG volvulus while performing fetal sonography [6]. Accordingly, early diagnosis of the maternal MG volvulus (usually, during the third trimester) also relies on thorough clinical evaluation and proper use of radiology [23].

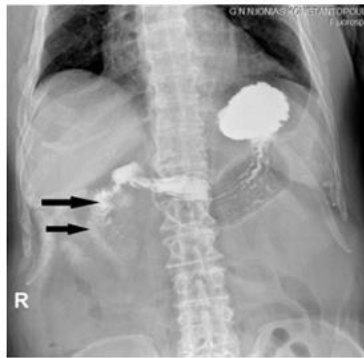
The hesitation to pursue radiological imaging during pregnancy for the risk of radiation exposure is often the main obstacle in achieving a definite diagnosis [23]. The maximum radiation that a fetus can be safely exposed to is 10 rads, but, currently no single diagnostic study exceeds 5 rads [23,27]. U/S (first line) and MRI have been reported to be safe and valuable in pregnancy, with no associated risks to the fetus [7,9,23,27]. However, modalities that rely on ionizing radiation, such as the abdominal X-ray (i.e., in pregnant woman with MG volvulus) and (low dose) CT scan have been safely used [8,9,23].

U/S in fetal MG volvulus may reveal proximal bowel obstruction with a closed loop and other dilated bowel loops, anatomic abnormalities of the SM vessels and the duodenum, and indirect signs, as well, such as polyhydramnios, ascites, meconium pseudocyst, and decreased fetal movements [4,6,7,19]. The hallmark “whirlpool sign” refers to a coiled configuration due to the torsion of the mesenteric root, and the superior mesenteric vein (SMV) wrapped around the SMA axis [7,19]. It is highly suggestive of MG volvulus; some studies in adults report sensitivity and specificity of 92% and 100% respectively [19]. The “coffee-bean sign” may occur in fetal MG volvulus cases with “closed loop bowel obstruction”, and refers to the coffee-bean-like appearance of the closely approximated ahaustral gas-filled colon; it must be differentiated from the severe meconium obstruction with intestinal torsion [6,7]. On U/S, the presence of “fluid-fluid level” within dilated bowel loops may improve the diagnostic accuracy, and on Doppler study the increased peak systolic velocity of the middle cerebral artery, especially with dilated bowel loops, ascites and polyhydramnios, can be indicative of fetal anemia [6].

Abdominal X-rays in childhood and adulthood may usually show evidence of dilated bowel loops displaced on the right side (possibly, with gastric distension) and multiple air-fluid levels [4,7,14,16,23]. Less commonly, MG volvulus in pediatric population or adults may be associated with signs of duodenal obstruction [4]. Abdominal X-rays can thus be helpful, but not always (Figure 1 a,b).



**Figure: 1a**



**Figure: 1b**

**Figure 1:** a) Abdominal X-rays: Dilated colon and air-fluid levels (arrows) in 50 year-old male with MGM-MG volvulus. b) Postoperative Gastrografin® UGI series in 70 year-old male with acute bowel obstruction due to MG volvulus on underlying MGM and rectosigmoid tumour; arrows indicate coexistent malformed right-sided duodenum (without 3rd and 4th portion) and right-sided proximal jejunum. Both were operated.

Abdominal CT scan for MG volvulus secondary to MGM in pediatric population and adults is the confirmatory study of choice, as it allows for identification of the highly suggestive “whirlpool sign” of the twisted mesentery and the wrapped SMV, rotated clockwise around the SMA, and the “bean-like

appearance” of the obstructed proximal jejunum [4,8,14,16,19,28]. SMA/SMV orientation abnormalities (inversion or vertical positioning), can be associated with positional anomalies of the duodenum and the undeveloped pancreas [4,5,13,28]. The CT findings may include dilated loops of small bowel sequestered on the right abdominal side, interloop ascites, and dilated right colon on the left, with the distal colon collapsed [14,19]. An ominous sign is the presence of intestinal ischaemia or necrosis [10]. Rarely, a right hepatodiaphragmatic entrapment of bowel (“Chilaiditi syndrome”-like) can be seen [16]. Contrast-enhanced CT scan has been evaluated to be effective in diagnosing MGM in adults with diagnostic rates between 80% and 97% [5,29]. Reviewing relative data, Butterworth WA and Butterworth JW [13] found that the diagnosis of acutely presented MGM was predominantly made via CT scan (67%), but also via U/S (15%) and intraoperatively (18%). However, in emergent cases, there may have been findings overlooked or missed by the preoperative clinical and imaging examination (Figure 2a-c).

In the evaluation of children with clinically suspected MGM, apart from the use of X-rays and U/S, the Gastrografin® upper GI (UGI) series have a place [3,4,8,11,30]. In the series of 22 patients (15 with MG volvulus) of Torres and Ziegler [3], UGI series were adequately sensitive (95%) and accurate (86%). With this modality, the “corkscrew appearance” of the duodeno-jejunal junction and its deviation to the left of the vertebral body suggest MGM [4]. As for the occasional dysmotility condition of the colon, it can be evaluated in symptomatic MGM cases with delayed orocaecal and colonic transit times [31].

Finally, emergency exploratory laparotomy can establish the intraoperative diagnosis in complicated or obscure cases [16]. Various expected and unexpected abdominal findings may be encountered (Figure 3a-c).

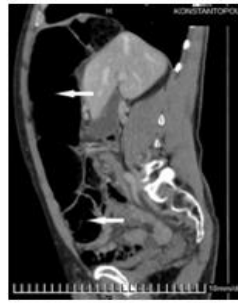
## Treatment

Despite the fact that several investigators advocate surgery in all patients with a diagnosis of MGM regardless of symptoms or age, there is a general agreement that the surgical intervention is the treatment of choice only for complicated MGM cases [12,16]. In these cases, MGM patients with acute onset of symptoms may already have haemodynamic instability with sepsis from bowel ischaemia or perforation or complete refractory obstruction, or they are in high risk to have it in the next hours [10,17,18,22]. What is primarily required in each emergent case is a quick clinical assessment and a laboratory work-up, and, for the unstable patient, immediate simultaneous intense fluid resuscitation and haemodynamic support to correct dehydration and electrolyte/metabolic disturbances (acidosis), nasogastric decompression and administration of broad-spectrum antibiotics [4,10,16,23]. The operation is performed openly due to the

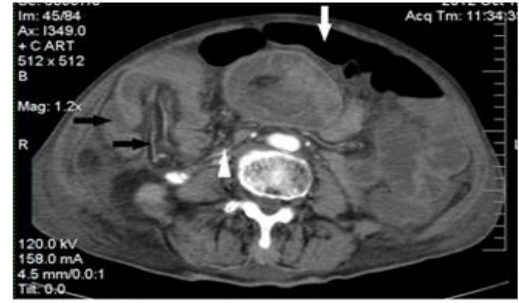
existent significant bowel distension, the altered anatomy, and concern for potential bowel injury with a laparoscopic approach [12,19,26].



**Figure: 2a**



**Figure: 2b**

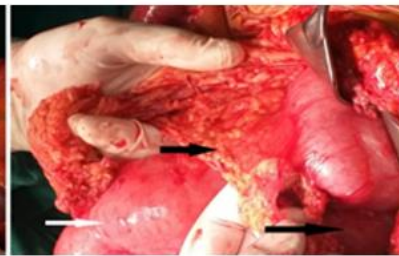


**Figure: 2c**

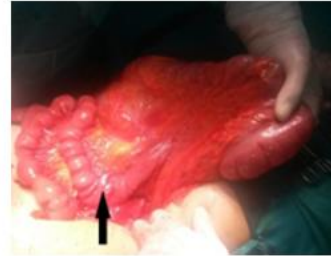
**Figure 2:** Abdominal CT scan: Complete obstruction with dilated colon (white arrows) in a 52 year-old male (a-axial, b-sagittal: closed-loop obstruction), and a 88 year-old female (c) with MGM-MG volvulus; arrowheads show the “whirlpool sign”; coexistent ICI (c) is indicated with black arrows. Both were operated.



**Figure: 3a**



**Figure: 3b**



**Figure: 3c**

**Figure 3:** Intraoperative findings of complicated MGM-MG volvulus with dilated/unfixed colon in a 50 year-old male (a: white arrow for right colon, black arrows for omentum and bands), in a 42 year-old male (b-after devolvulation : arrow for sequestered small bowel in the right side), and in a 46 year-old female (c: arrows for ileum and peritoneal bands).

However, in the acute setting, surgeons are occasionally confronted with underlying pathology with limited or lacking preoperative information, or unexpected findings, and are forced to use a treatment option with little evidence [25]. This is particularly true for complicated MGM cases.

### Treatment recommendations for neonates, infants and young children

The standard of care for neonates and infants suffering from symptomatic or complicated MGM (duodenal obstruction, MG volvulus) is the Ladd’s surgical procedure, originally proposed by William Ladd on 1936 [3-7,16,23,31]. In the vast majority of cases, characterized by absence of bowel necrosis, it involves: (i) counter-clockwise detorsion of the volvulised bowel (if MG volvulus is the case) with full inspection of the mesenteric root; (ii) division of Ladd’s bands and other congenital peritoneal adhesions (especially, around the SMA) to relieve obstruction and straighten the duodenum along the right abdominal gutter; (iii) broadening of the narrow mesenteric root, and re-orientation of the small bowel on the right hemiabdomen and the colon on the left side; and (iv) prophylactic appendectomy (usually), to prevent future diagnostic confusion in the setting of acute appendicitis [3-

6,13,14,23,31]. The rare case of intrauterine fetal MG volvulus mandates rapid and multidisciplinary management including double surgical intervention, an emergent caesarian section on the mother for delivering of the neonate, and an emergent laparotomy on the neonate for detorsion of its twisted bowel and prevention of intestinal necrosis [6,7,23]. Conclusively, the classic Ladd’s technique, which is not a true correction procedure, aims primarily to rescue newborns with complicated MGM [31]. In the absence of severe associated anomalies, the isolated MG volvulus has a favourable prognosis postoperatively [6].

The management of children with asymptomatic or minimally symptomatic or incidental MGM remains controversial [12,14,32]. The potential to develop sudden onset of acute complications in asymptomatic patients always exists at any age, thus, proponents of a prophylactic Ladd’s procedure site that the perioperative risk associated with an elective surgery is far lower when compared to an emergent one [12]. However, the traditional thought is that the asymptomatic MGM diagnosed after two years of life poses minimal risk. Avoiding surgery in older asymptomatic children and adults is also justified by the dramatically declining risk of presenting patients with volvulus with advancing age [12]. Moreover, it has been reported that,

correcting asymptomatic MGM beyond the age of 20 years is ineffective and probably harmful [4]. The 2015 American Pediatric Surgical Association (APSA) outcomes and evidence-based practice committee concluded that there is minimal evidence (Level 4) and recommendation (Grade D) to support screening, and recommended observation for older children with asymptomatic MGM [32]. An issue of concern is the recurrence of MG volvulus after the index surgery [5,9,26]. Many investigators suggest that the recurrence rate among children is low, and a pexy of bowels may constitute an unnecessary addition [14]. The 1980's Stauffer's series [33] with 77 children, who had undergone a Ladd's procedure with or without pexy, showed a trend for less reoperation in the group with pexy, but no significant difference in the recurrence rate between the two groups. However, the recurrence rate among patients who underwent Ladd's procedure alone in the adulthood is much higher than the one reported in children, estimated in some series as high as 16% [9] or even 18% [8]. Advocators of a complementary fixation during the index surgery or when treating recurrence have used various techniques, such as suturing of the posterior duodenum to the right renal fascia (duodenopexy), anchoring the caecum to the left abdominal wall (caecopexy), or some fixation of the small bowel or its mesentery (enteropexy, mesentericopexy) [14,19,26,31].

### **Treatment recommendations for emergent patients beyond infancy until adulthood**

In the emergency setting for older ages, the surgical bowel detorsion alone is not an option, since it is associated with a high risk of recurrence (20-75% in cases of right colon/caecal volvulus), besides the fact that bowel excision for ischaemic necrosis and correction of coexistent pathology are inevitably required [11,16]. Instead, some elements of the Ladd's method should complete the required operation, i.e., the division of Ladd's and thick peritoneal bands, the taken down of the "ligament" of Treitz with the creation of a "Treitz neo-ligament", and the straightening of duodenum to the right [5,11,15,19,23,31]. The type of the surgical operation in complicated cases is determined by the individualized variant of the developmental defect, the intraoperative situation of the bowel, and the patient's status for surgery [8,10,18,21]. It is crucial for the operating surgeon to recognize associated abdominal vascular and other coexistent anomalies, and tailor the surgery accordingly [4,10,31]. However, there remains much debate regarding the proper management for specific forms of primary MG volvulus in these ages [19,31].

An issue of concern is the place of colonoscopic devolvulation in complicated cases with RCV obstruction that is associated with non-gangrenous colon. In contrast to its application in sigmoid volvulus cases, this non-operative relief method is rarely used in

complicated MGM cases, since it is rarely successful and subsequent revolvulation often occurs, leading to a high risk of perforation [18,29,34]. However, it could sometimes convert an emergent procedure in a debilitated and poorly prepared patient to a semi-elective one, providing temporary mechanical detorsion and decompression of the bowel [18,34].

For the emergent stable patient, beyond the early period of life, who responds well to resuscitation, who has no risk factors, and who has bowel ischaemic necrosis or perforation or not, the optimal surgical management entails open right hemicolectomy with primary ileotransverse anastomosis for MG volvulus, and the rarest case of ICI on underlying MGM [10,18,21,34]. Prerequisites for this type of surgery are achieving viable and healthy bowel ends and the absence of adverse factors in anastomotic healing (i.e., malnutrition, steroid use, bowel edema) [10,18,22]. If the patient is at high risk for anastomotic leak or was preoperatively assessed with poor rectal tone or incontinent sphincter (elderly), then resection with creation of ileostomy remains a viable option [18,22,31]. Extended large bowel resection or, instead, segmental bowel resection and colopexy have also been reported in specific cases [10]. Non-resectional approaches in the setting of viable colon after devolvulation, such as the colopexy or combination of pexy and caecostomy, have been used as suboptimal surgical options but life saving measures on debilitated-malnourished patients; reportedly, detorsion alone is associated with recurrence rate as high as 75%, and detorsion with colopexy is associated with a recurrence rate up to 40% and a mortality up to 18% [16,18,22,31,33]. Notably, the presence or coexistence of ICI mandates that the resection should achieve negative oncological bowel margins [17,18,21].

On the other hand, for the emergent unstable patient with sepsis from bowel ischaemia or perforation, or complete obstruction (strangulation), characterized by one or more of metabolic acidosis, hypoxia, sustained hypotension and coagulopathy, there is no time for full resuscitation and optimization [18]. Reversal of haemodynamic instability will not occur unless bowel obstruction has been resolved. This patient in extremis should undergo simultaneous resuscitation and transfer to the operating room [10,18,34]. Damage control surgery is the option, involving rapid stapled in situ resection of the volvulised (rarely, intussuscepted) gangrenous bowel segments and mesentery (i.e., right hemicolectomy), control of spillage and lavage/debridement, proximal bowel diversion (usually), and temporary abdominal closure [10,13,18,34]. Intentionally, conserving the intestines in case of uncertainty is considered as a valid option instead of extended intestinal resection, especially in young patients [4]. This is a very important issue.

The choice of a technique of temporary closure in the case of significant bowel distension or when bowel of questionable viability was left in situ, is another issue of concern. For this case,

particularly in adults, a laparostomy dressing, an absorbable mesh (i.e., vicryl mesh) or a BOGOTA bag may be applied [4,13]. These patients are better closely attended in the surgical intensive care unit, so as to promptly recognize and treat every exacerbation of metabolic /lactic acidosis, decrease in urinary output and worsening of laboratory examinations, indicating tissue reperfusion injury or that the bowel segment in uncertainty has not recovered or is necrotizing [4]. Based on the degree of response to resuscitation after index surgery, a return to the operating room within 24-48 hours for re-look (s) is required [18,34]. Bowel reconstruction in cases with healthy bowel, or, instead, complementary excision when bowel necrosis is evident, should accordingly be performed [4,13].

The postoperative complications, minor in most of the cases, are associated with the present developmental anomalies, the type of surgery performed, and the patient's physical status. They include wound infection, adhesive ileus, delayed gastric emptying, and recurrence [8]. Unfortunately, complicated MGM that is associated with excessive bowel necrosis leads to extended bowel resection, either during the index surgery or during re-operation; patients with "short bowel syndrome" require close and continuous caring for malnutrition (in need of lifelong parenteral nutrition), electrolyte disturbances, immune deficiency, and even sepsis [23].

In the emergency setting, the role of laparoscopic intervention for complicated MGM remains limited. Theoretically, in symptomatic but stable patients with absence of bowel necrosis or excessive bowel distension, the Ladd's procedure can be performed laparoscopically, gaining less postoperative pain and shorter time to full enteral feeding and hospital stay. In practice, the physical status of these few patients undergoing laparoscopy for MGM is markedly better than those who undergo laparotomy; nonetheless, up to one third of the laparoscopic procedures are eventually converted to open surgeries [4,12,16,35,36]. There is also evidence of an increased risk of volvulus recurrence with the laparoscopic approach in children due to the poor formation of post-operative adhesions [12,36]. However, laparoscopy for MGM with bowel obstruction has very rarely been reported, probably due to perceived difficulties with the use of laparoscopic tools and the rarity of this condition [15,34]. Prospective randomized studies with long follow-up are required to evaluate the true efficacy of the laparoscopic approach.

## Conclusion

MGM refers to various congenital anomalies of the rotation and fixation of the midgut, often resulting in the creation of an unfixed, hypermobile, and displaced caecocolon, prone to twisting, and the presence of abnormal peritoneal bands compressing the duodenum or the bowel. Acute presentations of MGM are associated with bowel obstruction with risk of bowel

necrosis, a dreadful complication that is more commonly and easily diagnosed early in the life, while it occurs rarely and diagnosed with delay in older ages. Acute cases mandate appropriate resuscitation and emergent surgery to relieve bowel obstruction (Ladd's procedure in early life cases with no necrosis and bowel resection for necrosis or in older cases), and to repair coexistent congenital aberrations. Higher mortality rates are seen in cases with acute onset of MG volvulus, delayed diagnosis, or presence of bowel necrosis. High index of clinical suspicion for timely diagnosis and surgery, sufficient knowledge of the normal and altered anatomy, coupled with understanding of the mechanisms of disturbance in complicated cases, and familiarity with basic surgical techniques should guide practitioners to handle difficult unexpected findings.

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## Conflict of interest

The author has no conflicts of interest to declare.

## Ethical Approval informed Consent

Written informed consents were obtained from the patients whose images are used for presentation in Congresses or publication in Medical Magazines.

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