

Contents lists available at [ScienceDirect](https://www.sciencedirect.com)

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Midgut volvulus caused by intestinal malrotation; A rare cause of acute abdomen in adults. Case report

Fernando Perez Galaz, Karen Moedano Rico, Felix Alejandro Pérez Tristán, Alejandro Acuña Macouzet, Marcos Jafif Cojab*

Department of Surgery Hospital Angeles Lomas, Edo. Mex, 52763, Mexico

ARTICLE INFO

Article history:

Received 31 May 2020

Received in revised form 14 July 2020

Accepted 14 July 2020

Available online 18 July 2020

Keywords:

Intestinal
Ischemia
Occlusion
Ladd's bands
Malrotation

ABSTRACT

INTRODUCTION: Acute abdomen due to midgut volvulus with intestinal malrotation is rare event with only only few cases in the literature Butterworth et al. (2018). Intestinal malrotation presented in the adulthood is reported in 0.2–0.5 %, of cases; with only 15 % of them presenting as midgut volvulus Butterworth et al. (2018).

Intestinal malrotation is caused by an alteration in embryonic development between 10–12 weeks of gestation. The main alteration is anomalous position of the bowel with the small intestine residing on the right side of the abdomen, while the colon and cecum remain on the left side due to malposition of the Treitz ligament. Additionally, the ascending colon remains attached to the abdominal wall by fibrous peritoneal bands known as Ladd bands, this being a cause of midgut volvulus and intestinal obstruction. **PRESENTATION OF CASE:** We present a 25-year-old male with failure to thrive who arrives at the ED with clinical signs and symptoms of intestinal occlusion and acute abdomen, initial resuscitation is made in ED and is transferred to OR, an exploratory laparoscopy evidencing intestinal malrotation with cecal volvulus and a Ladd procedure is made openly without PO complications.

DISCUSSION: The gold standard for diagnosis of intestinal malrotation is the upper gastrointestinal series. However, in patients with acute abdomen associated with this pathology where is suspected intestinal ischemia and hemodynamic instability, it is essential that an emergency laparotomy be performed.

CONCLUSIONS: The diagnosis of intestinal malrotation is difficult, since many patients are asymptomatic in adulthood or present with variable GI symptoms. Therefore, it is imperative to have a high index of suspicion in patients with compatible clinical characteristics in order to perform the best therapy in time and manner.

© 2020 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Intestinal malrotation is the product of a well-known alteration in the normal embryonic gut development that occurs between 10–12 weeks of gestation, since the consequences associated with this pathology can cause severe morbidity or mortality, it is imperative to understand the embryology, anatomy, diagnostic criteria and treatment [1,2].

An incidence of 1 in 500 newborns is estimated, however symptomatic malrotation in neonates occurs with a frequency of about 1 in 6000 live births, usually presenting in the first month of life in 64–80 % of patients as volvulus or intestinal obstruction [3]. However, more recent studies have shown 30 % presentation at one

month of age, 58 % before one year of age and 75 % before five years of age [13].

Adult presentation is extremely rare with an incidence of 0.2 % [6]. Therefore the diagnosis is often delayed with a significant increase of morbidity, some patients may be asymptomatic throughout life or presenting symptoms in adulthood insidiously in the postprandial period such as; intermittent biliary vomiting (30 %), intermittent abdominal pain (20 %), failure to thrive, oral intolerance, chronic diarrhea, and malabsorption. These symptoms are usually present for a period of about 6 months before the diagnosis is made [3,4].

More importantly midgut volvulus is a rare presentation of cause acute abdomen in the adult population with only 93 cases reported in literature [14]; these patients present severe abdominal pain, signs of intestinal obstruction, hematemesis or hematochezia and in some occasions with hemodynamic instability [5,6,14].

* Corresponding author at: Department of Surgery, Vialidad de la barranca S/N, Hospital Angeles Lomas, Huixquilucan Edo. Mexico, 52763, Mexico.
E-mail address: drmarcosjafif@gmail.com (M. Jafif Cojab).



Fig. 1. Abdominal X-Ray showing small bowel occlusion pattern with multiple air fluid levels, small intestine with predominance in the left hemi-abdomen and absence of air in the RLQ suggestive of malrotation.

2. Case report

A 25 year old male patient presented to the Emergency department with the chief complaint of abdominal pain which was sudden in nature, 8/10 in the scale of pain with irradiation to the RLQ, the pain was accompanied by nausea and multiple episodes of vomiting gastric content, the patient also referred inability to pass stool or flatus in the last 24 h.

Physical examination was remarkable for appearance age less than chronological, small stature and low weight

On physical examination vital signs BP: 110/67 mmHg, HR: 101 T 36.7 abdominal exploration was remarkable for absence of peristaltic sounds, pain to palpation in the RLQ with rebound sign.

Laboratory studies did reveal leukocytosis and electrolyte imbalance with mild hypokalemia, severe volume depletion was noted, abdominal X-ray was ordered revealing a small bowel occlusion pattern and absence of gas in the RLQ suggestive of malrotation (Fig. 1).

Nasogastric tube was inserted and 250cc of gastric content were drained without relief of the distention and pain.

Small bowel occlusion diagnosis was made and no further images were made.

Fluid resuscitation was started and broad-spectrum antibiotics were given.

We proceed with an exploratory laparoscopy, at entering the peritoneal cavity bowel distention was remarkable with distal ileum collapse, midgut volvulus was seen and multiple Ladd's bands attaching the ascending colon to the duodenum, we decided to convert the procedure to a laparotomy due to the difficulty in handling distended bowel loops in a small abdominal cavity.

At the exploratory laparotomy we made a Ladd procedure beginning with the evisceration of the bowel, afterwards we untwisted the volvulated midgut, after inspecting the bowel for few minutes we determined no viability of the entire ascending colon, we



Fig. 2. Intra-operative image showing dilated small bowel loops and midgut volvulus due to multiple bands attaching the ascending colon to the small intestine caused by intestinal malrotation.

made band lysis we proceed with a right hemi-colectomy due to ischemic section of the bowel, we made an ileo-colic anastomosis with a linear stapler and finished the procedure without any incident, surgical specimen was sent to pathology (Figs. 2–6).

Post-operative recovery was uneventful returning to oral feeding in the first 24 h; bowel movements were present on post-operative day 2 and discharged on post-operative day 4.

3. Discussion

The term poor bowel rotation was described by Ladd [2] in 1936 and can be defined as: a congenital anomaly of intestinal rotation and fixation during the development of the fetus.

Changes in intestinal embryological development can occur in any of its phases and can be grouped according to the corresponding stage of development [2,7].

In the first phase or herniation phase, which occurs during the first 8 weeks of gestation, the intestine protrudes through the yolk sac, due to rapid intestinal growth, particularly of the ileum, and rotate 90 degrees counterclockwise, failure to rotate at this time will produce an omphalocele.

In the second phase / Return to the abdomen, during week 10 and 11 of gestation, the abdominal cavity enlarges, the intestine retracts from the umbilical cord and returns to the abdomen, the duodenojejunal loop is the first to return and subsequently rotates counterclockwise 180-degrees (Total 270-degrees) on the axis of the artery superior mesenteric. Normal rotation and fixation result in a wide mesenteric base that extends from the ileocecal valve to the ligament of Treitz. In this phase the abnormal rotation, due to failure to complete the 180-degree rotation will result in a narrow mesenteric base and a vascular pedicle susceptible to volvulus. The resulting spectrum of alterations include; no rotation and incomplete rotation [1,7].

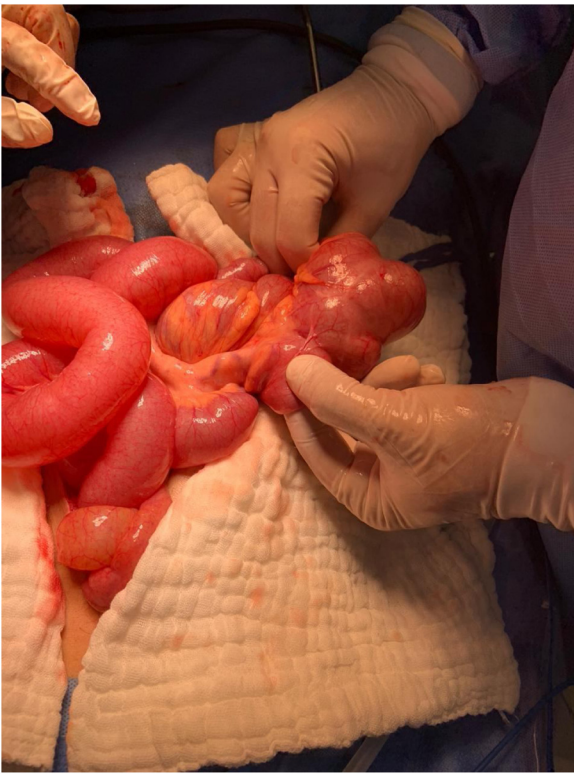


Fig. 3. Intra-operative image showing bands attaching the small bowel to the ascending colon.



Fig. 5. Intra operative image showing midgut volvulus with twisting of the small bowel around the ascending colon and cecum.



Fig. 4. Intra-operative image showing band of fibrous tissue attaching the small bowel to the right lateral wall.



Fig. 6. Surgical specimen showing the terminal ileum, cecum and ascending colon, multiple bands are still present in the specimen.

In non-rotation the duodenum descends to the right side of the superior mesenteric artery, resulting in a small intestine completely in the right hemiabdomen and colon in the left hemiabdomen. This condition is usually asymptomatic [1].

Incomplete rotation or malrotation, occurs when rotation is arrested at this stage. The duodenal loop lacks 90-degrees of the total 270-degrees, and the ceco-colic loop lacks 180-degrees of its normal rotation. As a result, the cecum remains on the left side due to the lack of development of the Treitz ligament, however, it remains attached to the right abdominal wall by fibrous peritoneal bands known as Ladd bands [2], these generally surround the duodenum producing the symptoms of intermittent intestinal obstruction [1,3,7].

The narrow mesenteric base and vascular pedicle makes the small bowel high risk for volvulus [8].

In the third or fixation phase, which occurs from week 12 to birth, failure results in a mobile cecum, sub-hepatic cecum, or retro-cecal appendix. On other occasions there may also be failure of intestinal mesenteric fixation, which conditions susceptibility to volvulus [1].

The diagnosis of intestinal malrotation should be suspected in adults who have the clinical characteristics described above, or who have abnormal findings in imaging studies performed for other reasons. The *gold standard* for diagnosis in both pediatric and adult patients is the upper gastrointestinal series, since it is the tool that best visualizes the normal path of the duodenum; a duodenal obstruction with a “beaked” appearance is suggestive of volvulus or the classic appearance of corkscrews in the first jejunal loops [8]. However, in patients with an acute abdomen associated with this pathology, in whom intestinal ischemia is suspected, it is essential that they be taken to the operating room without the need for any of image study [8].

Another useful diagnostic tool is abdominal computed tomography, which identifies rotation patterns that could be associated with particular complications [9]. However, in more recent series it has been shown to have the same sensitivity and specificity as the upper gastrointestinal series [10].

Abdominal ultrasound is not indicated to make the diagnosis since a negative result does not exclude the presence of malrotation; its importance should be highlighted since it can be diagnosed incidentally in a study carried out for other reasons [12].

The management of these patients depends on their initial presentation, in asymptomatic patients or with gastrointestinal manifestations that are not life threatening, a Ladd procedure can be performed electively.

The Ladd procedure is the surgical therapy of choice, it can be performed openly or laparoscopically since there are no significant differences in the rates of complications, need for reoperation or the persistence of symptoms [6].

The procedure described by William Ladd in 1936 continues to be the therapy used in these cases, and consists of 5 steps; identification and distortion of the volvulus counterclockwise, division of the Ladd bands, division of the inter-mesenteric bands (fibrous bands between non-cecal intestinal loops and the duodenum) for widening of the mesenteric base and appendectomy, to end the procedure the intestine is placed in its anatomical normal position. Additionally, any frankly necrotic bowel should be resected and an anastomosis performed. If there is suspicion of the viability of the intestinal loops, it is recommended to perform a “second look” surgery within 24–48 h [12].

Oral feeding can be safely started within 24 h after surgery preventing post-operative complications [15].

Patients with acute abdomen secondary to intestinal volvulus and ischemia require emergency laparotomy, however, adequate resuscitation is required in the preoperative period with fluids, the placement of a nasogastric tube and the initiation of broad-

spectrum antibiotics; These measures should not delay the start of surgery and should be done while the patient is being transferred to the operating room. On the other hand, it should be noted that in many cases, such as this one, the diagnosis was made intraoperatively since the hemodynamic instability of the patient made immediate surgical management essential.

4. Conclusions

Midgut volvulus due to intestinal malrotation is a rare cause of acute abdomen in the adult population. The diagnosis of intestinal malrotation is delayed in some cases, since many adult patients are asymptomatic or present variable gastrointestinal disturbances long before reaching the diagnosis. Since the initial presentation of some patients is in the form of an acute abdomen secondary to intestinal volvulus and compromise of intestinal viability, it is essential that those patients undergo an exploratory laparoscopy/laparotomy, in many cases omitting the pertinent diagnostic studies such as the upper gastroduodenal series or computed tomography.

Due to the potential catastrophic outcome of this pathology, it is imperative to have a high index of suspicion in patients with compatible clinical characteristics in order to perform the best therapy in time and manner.

Declaration of Competing Interest

There was no conflict of interest.

Funding

There was no sponsorship for this study.

Ethical approval

There was no need for ethical approval.

Consent

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

Author contribution

Fernando Perez Galaz MD FACS: Study design
 Karen Modeano Rico MD: Study design
 Felix Alejandro Perez Trsitán MD: Data collection, data analysis, writing the paper
 Alejandro Acuña Macouzet MD: Data collection, data analysis
 Marcos; Jafif Cojab MD: Data collection, data analysis, writing the paper

Registration of research studies

None.

Guarantor

Fernando; Perez Galaz MD FACS.

Acknowledgement

We acknowledge Oscar Cervantes Gutierrez MD and Gustavo Cervantes Millan MD for their valuable contribution for this case report.¹

References

- [1] A.M. Torres, M.M. Ziegler, Malrotation of the intestine, *World J. Surg.* 17 (1993) 326–331.
- [2] W.E. Ladd, Surgical diseases of the alimentary tract in infants, *N. Engl. J. Med.* 215 (705) (2020) 1936.
- [3] B.W. Haak, S.T. Bodewitz, C.F. Kuijper, L.M. de Widt-Levert, Intestinal malrotation and volvulus in adult life, *Int. J. Surg. Case Rep.* 5 (2014) 259–261.
- [4] R. Yanez, L. Spitz, Intestinal malrotation presenting outside the neonatal period, *Arch. Dis. Child.* 61 (1986) 682–685.
- [5] H.B. Devlin, R.S. Williams, J.W. Pierce, Presentation of midgut malrotation in adults, *Br. Med. J.* 1 (1968) 803–807.
- [6] Adrian T. Fung, et al., Malrotation with midgut volvulus in an adult: a case report and review of the literature, *J. Surg. Case Rep.* 5 (2017).
- [7] E. Hernando-Almudí, R. Cerdán-Pascual, C. Vallejo-Bernad, J. Martín-Cuartero, M. Sánchez-Rubio, M. Casamayor-Franco, Malrotación intestinal en adulto asociada a vólvulo intestinal, *Cirugía y Cirujanos* 85 (2017) 424–427.
- [8] H. Kotobi, V. Tan, J. Lefèvre, F. Duramé, G. Audry, Y. Parc, Total midgut volvulus in adults with intestinal malrotation. Report of eleven patients, *J. Visc. Surg.* 154 (3) (2017) 175–183.
- [9] B. Yang, W.H. Chen, X.F. Zhang, Z.R. Luo, Adult midgut malrotation: multi-detector computed tomography (MDCT) findings of 14 cases, *Jpn. J. Radiol.* 31 (5) (2013) 328–335.
- [10] E.T. Durkin, D.P. Lund, A.F. Shaaban, M.J. Schurr, S.M. Weber, Age-related differences in diagnosis and morbidity of intestinal malrotation, *J. Am. Coll. Surg.* 206 (2008) 658–663.
- [12] L.L. Frasier, G. Levenson, A. Gosain, J. Greenberg, Laparoscopic versus open Ladd's procedure for intestinal malrotation in adults, *Surg. Endosc.* 29 (6) (2015) 1598–1604.
- [13] J. Aboagye, S.D. Goldstein, J.H. Salazar, et al., Age at presentation of common pediatric surgical conditions: reexamining dogma, *J. Pediatr. Surg.* 49 (6) (2014) 995–999, <http://dx.doi.org/10.1016/j.jpedsurg.2014.01.039>.
- [14] W.A. Butterworth, J.W. Butterworth, An adult presentation of midgut volvulus secondary to intestinal malrotation: a case report and literature review, *Int. J. Surg. Case Rep.* (2018), <http://dx.doi.org/10.1016/j.ijscr.2018.07.007>.
- [15] G. Herbert, R. Perry, H.K. Andersen, et al., Early enteral nutrition within 24 hours of lower gastrointestinal surgery versus later commencement for length of hospital stay and postoperative complications, *Cochrane Database Syst. Rev.* 10 (10) (2018), <http://dx.doi.org/10.1002/14651858.CD004080.pub3>, CD004080. Published 2018 Oct 24.
- [16] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, SCARE Group, The PROCESS 2018 statement: Updating consensus preferred reporting Of CasE series in surgery (PROCESS) guidelines, *Int. J. Surg.* 60 (2018) 279–282.

Open Access

This article is published Open Access at [sciencedirect.com](https://www.sciencedirect.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited. This work has been reported in line with the SCARE criteria [16].