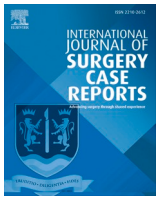
International Journal of Surgery Case Reports 91 (2022) 106750

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr



Case report

Variations of intestinal malrotation in adults: A case report of midgut volvulus and literature review for the surgeon

Tiago Ribeiro [a,*], Brittany Greene [a], Sean Bennett [a], Haytham Msallak [a], Paul Karanicolas [a,b]

[a] Department of Surgery, University of Toronto,149 College St, Toronto M5T 1P5, ON, Canada
[b] Sunnybrook Health Sciences Centre, 2075 Bayview Ave, Toronto M4N3M5, ON, Canada

ARTICLE INFO

Keywords:
Intestinal malrotation
Midgut volvulus
Adult
Case report

ABSTRACT

Introduction: Intestinal malrotation is a rare congenital abnormality occurring in 0.2–1% of the population. Adult presentations comprise only 0.2–0.5% of all cases leading to diagnostic challenges and worse outcomes in adults. We present a rare case of chronic/intermittent midgut volvulus with unique anatomic findings in an adult with intestinal malrotation.

Presentation of case: An 18-year-old Caucasian male presented to a community hospital with abdominal pain, nausea, and vomiting. He underwent a CT scan demonstrating concern for small bowel volvulus and subsequently underwent a negative exploratory laparotomy. He was discharged post-operatively with no identified etiology for his presentation. He subsequently had multiple presentations to the ED with recurrent symptoms, review of imaging led to concern for duodenal volvulus resulting in transfer to a tertiary hepatobiliary centre. Repeat CT scan two weeks following initial presentation was consistent with intestinal malrotation with midgut volvulus. Bloodwork was unremarkable and physical exam demonstrated normal vital signs with a tender epigastrium. He underwent an exploratory laparotomy with Ladd's procedure. Intra-operative findings included a midgut volvulus and uniquely positioned Ladd's bands to the transverse colon. Post-operatively he tolerated oral intake and was discharged with three-month follow-up.

Discussion: Adults with intestinal malrotation suffer from delays in diagnosis and management. In contrast to the neonatal population, adults often present with vague, or chronic symptoms, which obscures the diagnosis.

Conclusion: The increased morbidity and mortality observed in adults with intestinal malrotation highlights the need for surgeons to appreciate the challenges associated with this diagnosis in the adult population to ensure early recognition and management.

1. Introduction

Intestinal malrotation is a congenital abnormality occurring in 0.2%–1% of the population leading to a range of clinical challenges [1]. The term encompasses all abnormalities caused by rotational errors during embryonic development of the midgut. The majority of intestinal malrotation (60–80%) presents in the neonatal period as an acute surgical emergency secondary to midgut volvulus [2,3]. Failure in recognition, or management of this diagnosis may lead to significant sequelae including short gut syndrome, intra-abdominal sepsis and/or death [1]. The presentation beyond the neonatal period is more variable. Current literature suggests a decreasing incidence as patients age, and variable presentation with both acute and/or chronic symptoms [1,4–6]. Given the rarity of this diagnosis in adults, there remain many barriers to timely diagnosis and management including vague symptoms and provider inexperience. These barriers contribute to an increased morbidity and mortality observed with intestinal malrotation in adults [4,5,7].

We present here a unique case of an 18-year-old male with delayed management of intestinal malrotation presenting with acute on chronic symptoms, a chronic midgut volvulus and unique anatomic findings. A review of the literature of intestinal malrotation in adults will follow. This case report has been reported in line with the SCARE 2020 criteria [8].

* Corresponding author.
E-mail addresses: tiago.ribeiro@mail.utoronto.ca (T. Ribeiro), brittanydgreene@gmail.com (B. Greene), sean.bennett@mail.utoronto.ca (S. Bennett), haytham.msallak@mail.utoronto.ca (H. Msallak), Paul.Karanicolas@sunnybrook.ca (P. Karanicolas).

https://doi.org/10.1016/j.ijscr.2021.106750
Received 5 December 2021; Received in revised form 30 December 2021; Accepted 31 December 2021
Available online 4 January 2022

2. Presentation of case

A previously healthy 18-year-old Caucasian male presented to a community hospital with acute abdominal pain associated with nausea, vomiting, and a 3-month history of 22 kg weight loss. He was on no medications, had no known allergies, and no significant family history. Investigations included a CT scan concerning for small bowel volvulus leading to an exploratory laparotomy through a limited incision. The small bowel was run from the ileocecal valve to the ligament of Treitz finding engorged mesenteric veins but no signs of volvulus or ischemia. Post-operatively, his diet was advanced, and he was discharged. Shortly following discharge, he began to experience post-prandial abdominal pain associated with nausea and vomiting. This led to several emergency department presentations with ongoing diagnostic uncertainty in the setting of a recent negative laparotomy. Upon radiologist review of initial imaging, concern for possible duodenal volvulus arose leading to transfer to a tertiary hepatobiliary center.

Repeat CT scan, 2 weeks after initial presentation demonstrated several findings in keeping with intestinal malrotation including duodenal swirl, an obliterated SMV and numerous large venous collaterals (Fig. 1). The transverse colon was noted to cross the midline twice, an unexpected finding even in the context of intestinal malrotation (Fig. 1). Bloodwork was unremarkable. On exam the patient had normal vital signs and a soft abdomen with epigastric tenderness. The patient was consented for a laparotomy and Ladd's procedure, the standard of care for patients with intestinal malrotation. Intra-operative findings included a small bowel volvulus with mesenteric venous collateralization (Fig. 2). The duodenojejunal junction was in the right hemiabdomen with Ladd's bands uniquely placed coursing from the transverse colon to the right upper quadrant (Fig. 2). These findings were in keeping with intestinal malrotation with volvulus. The midgut was delivered into the operative field, and volvulus detorted through a counter-clockwise rotation. There were no signs of intestinal ischemia, which in conjunction with the engorged mesenteric vasculature was suggestive of a chronic, or intermittent midgut volvulus. The Ladd's bands were divided releasing the transverse colon from its attachments to the right lateral abdominal wall. Additional congenital bands were divided, an inversion appendectomy was performed, the mesentery was broadened, and the small bowel was placed free of tension in the right hemiabdomen and the large bowel in the left hemiabdomen.

Post-operatively his diet was advanced, and he was discharged on post-operative day 5. There were no complications or adverse events. Three months post-operatively he was tolerating oral intake with no pain. Post-operative CT scan to re-assess mesenteric vasculature demonstrated attenuation of the SMA and SMV with resolution of the swirl with expected post-operative arrangement of the bowels in a non-rotated configuration (Fig. 3).

3. Discussion

Intestinal malrotation is a rare diagnosis in adults with increased morbidity and mortality when compared to the pediatric population [4,9]. The worse outcomes in adulthood are multi-factorial. Contributing factors include the low incidence, varied clinical presentations, and uncertainty with ideal management [4,9]. Our case emphasizes the importance of considering intestinal malrotation in adults. Furthermore, the unique anatomic findings of chronic/intermittent volvulus with abnormal positioning of Ladd's bands highlight the possibility of anatomic variation resulting in a range of symptoms.

Normal midgut development is a complex process that requires 270° counter-clockwise rotation of the gut tube around the SMA axis [1,10]. This rotation usually occurs between the 4th–10th week of gestation creating a broad-based mesentery extending from the ligament of Treitz in the left upper quadrant, to the ileocecal valve in the right lower quadrant with retroperitoneal attachments to the duodenum and colon [1]. It is this broad-based mesentery that prevents volvulus in the normal anatomical configuration.

Intestinal malrotation encompasses a spectrum of clinical entities based on the anatomic configuration of the midgut after the rotational period is complete [10]. On one extreme of the spectrum is nonrotation, whereby the duodenum travels inferiorly on the right hemi abdomen, with all the small bowel located on the right side and colon on the left (Fig. 4) [1]. Classically, non-rotation presents in children with abdominal wall defects or congenital diaphragmatic hernia. On the other end of the spectrum, we can consider reverse rotation (Fig. 4). Reverse rotation, being the rarest form, results from clockwise rotation of the midgut in relation to the SMA. This leaves the transverse colon posterior to the SMA prone to extrinsic obstruction with the duodenum anterior to the SMA [1,10]. The remaining, and majority, of cases of intestinal malrotation involve incomplete rotation of the midgut creating a narrow

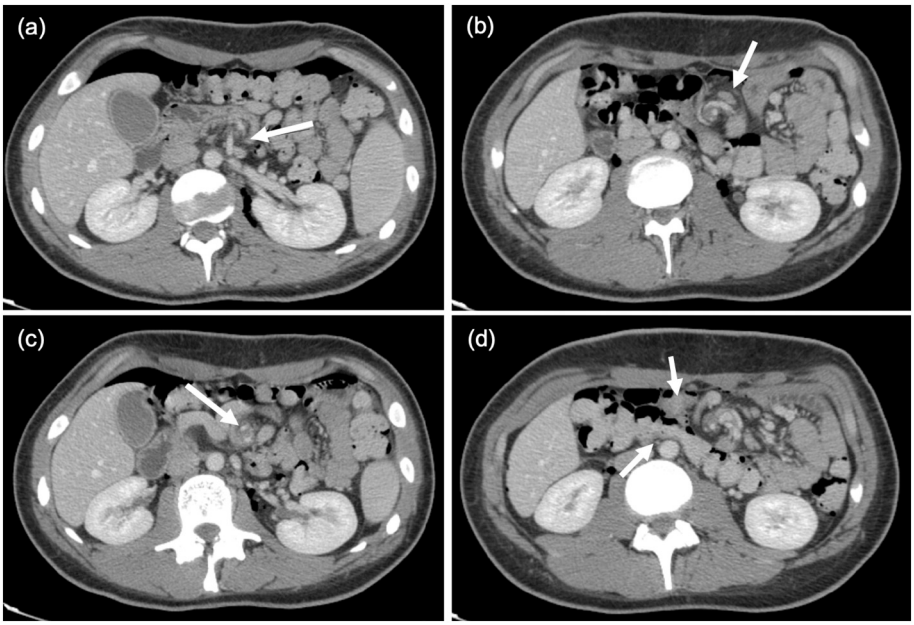


Fig. 1. Pre-operative CT scans venous phase demonstrating; (a, b, c) Duodenal swirl located left of the midline (d) transverse colon crossing midline twice.

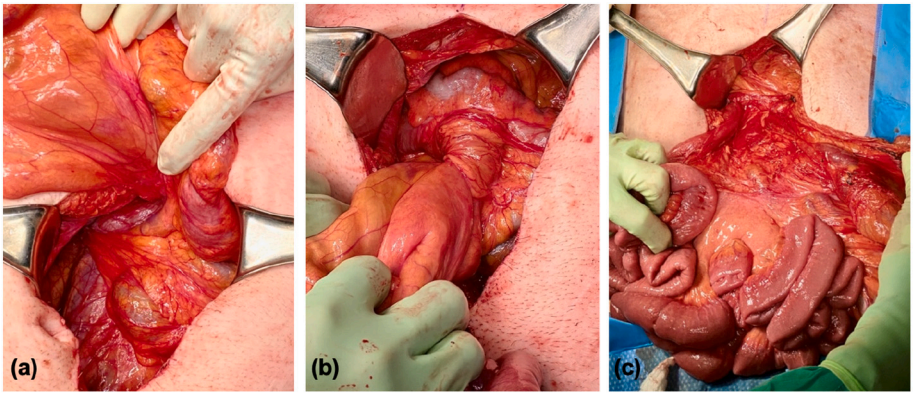


Fig. 2. Intra-operative photographs demonstrating; (a) Peritoneal (Ladd's) bands coursing from the transverse colon to the right lateral abdominal wall, (b) Midgut volvulus, (c) mesentery fanned out following lysis of bands and detorsion demonstrating engorged mesenteric veins.

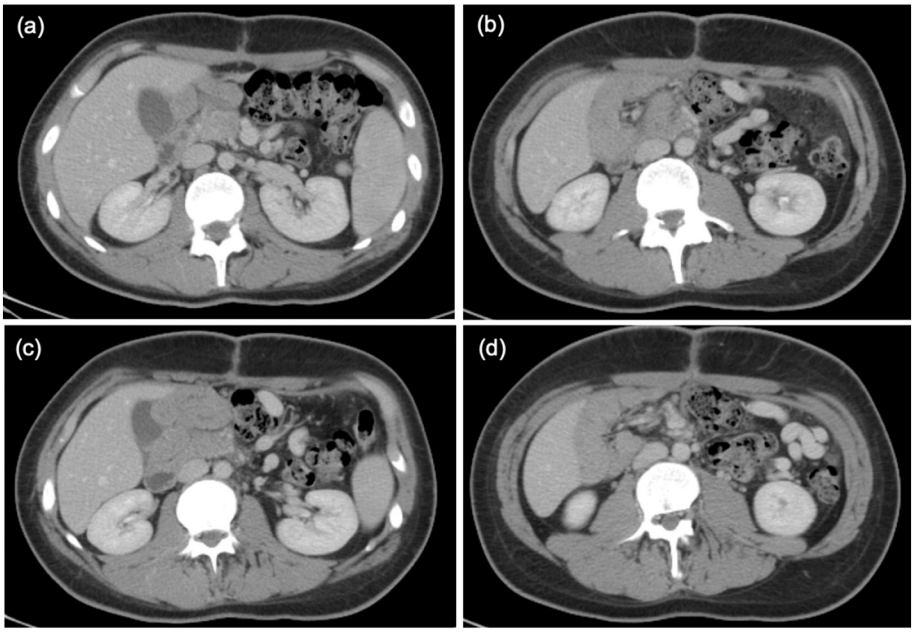


Fig. 3. Three months post-operative CT scans demonstrating resolution of duodenal swirl, small bowel located right of midline and large bowel located left of midline. SMV and SMA remain attenuated but no longer rotated.

vascular pedicle at risk of volvulus and lead to the formation of embryonically mispositioned adhesions, or peritoneal bands, known as Ladd's bands (Fig. 4) [1,10]. These bands classically fix the cecum to the right lateral abdominal wall in the upper abdomen which may lead to extrinsic obstruction of the duodenum [1]. The case reported above was incomplete rotation with unique findings. These include a transverse colon that crosses the midline twice, Ladd's bands fixing the transverse colon to the right lateral abdominal wall, and a chronic/intermittent volvulus (Fig. 4).

The current literature suggests that 90% of cases of intestinal malrotation present in the first year of life, the majority with acute bilious vomiting secondary to midgut volvulus [1,6,11]. Additional causes of the acute presentation include obstructing Ladd's bands and internal hernias [1]. Beyond the neonatal period, malrotation presents with decreasing frequency with both acute and chronic symptoms [4]. Although acute obstruction secondary to midgut volvulus or obstructing bands can occur in all age groups there is a tendency towards more obscure or chronic symptoms as a child ages [9,11,12].

In the adult population, intestinal malrotation presents similar trends as the older child with only 0.2–0.5% of all cases diagnosed in adulthood [9,13]. The presentation in adults is quite variable – presenting with symptoms that are acute, acute on chronic, chronic, or found incidentally [3,9,14]. Acute presentations of intestinal malrotation are less common with a retrospective review by Durkin et al. finding only about 20% of all adult cases present acutely [7]. The acute presentation in adults is usually secondary to volvulus leading to ischemia and eventual bowel necrosis with potentially fatal consequences [7,14,15]. In contrast to the pediatric population, adults who present acutely with intestinal malrotation have often experienced chronic symptoms which are only later attributed to their anatomic anomaly [4]. These chronic symptoms include intermittent or recurrent abdominal pain, vomiting, food intolerance and weight loss [4]. The diverse range of symptoms caused by intestinal malrotation in patients who have escaped the pediatric period results in diagnostic challenges for this rare diagnosis.

Given the heterogeneity of clinical presentations, appropriate imaging to investigate a patient's clinical presentation is imperative to diagnose intestinal malrotation and its complications in adults. CT scan has consistently been reliable in identifying several features consistent with malrotation including; whirlpool sign, SMA/SMV abnormalities, an underdeveloped pancreas or absence of the uncinate process, and

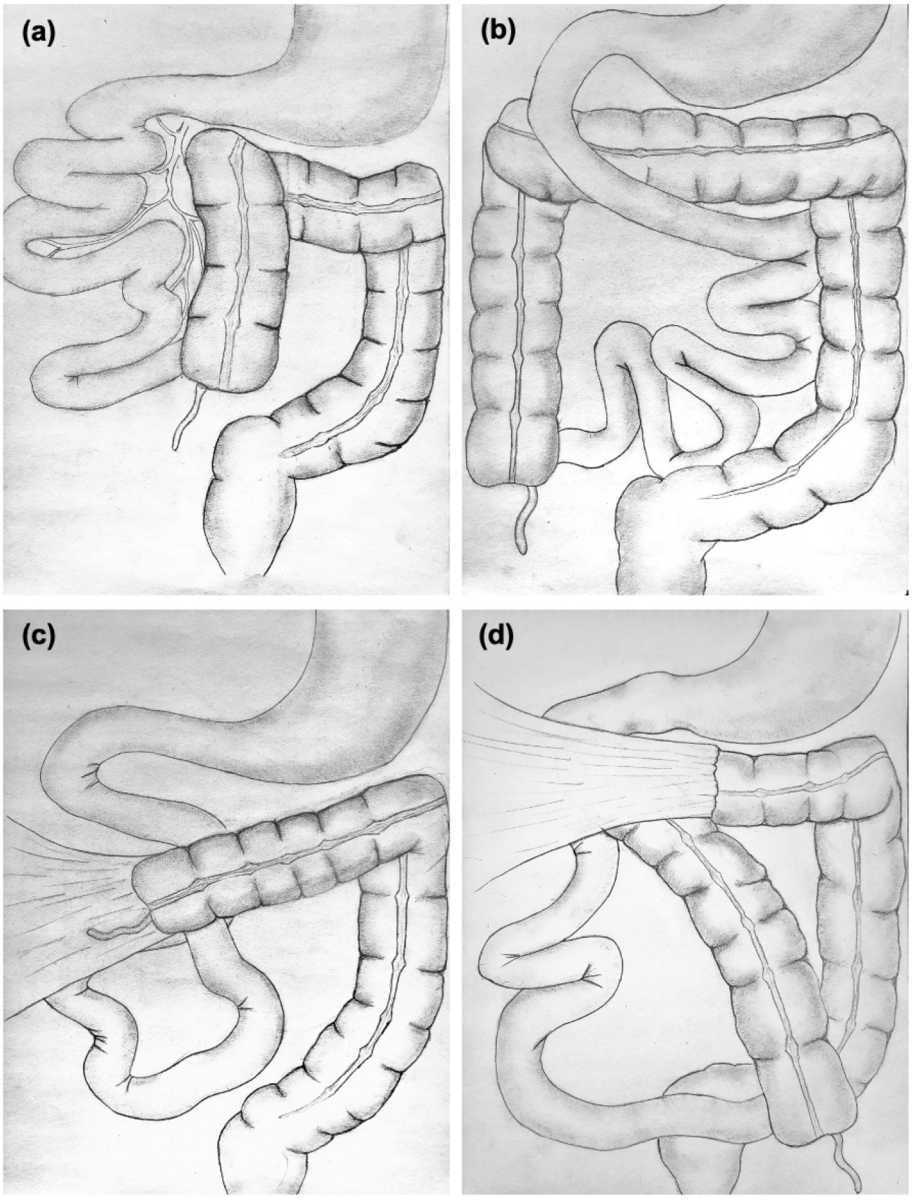


Fig. 4. Schematic diagrams of the classification of intestinal malrotation and our reported case. (a) nonrotation, (b) reverse rotation, (c) incomplete rotation, (d) case reported.

positional abnormalities of the duodenum, small intestine and cecum [4,5]. CT scan has been effective at diagnosing intestinal malrotation in adults with diagnostic rates between 80 and 97% [4,5,16].

The standard of care for patients presenting with symptomatic intestinal malrotation is the Ladd's procedure [12,14]. Ladd's procedure involves full inspection of the midgut structures, detorsion of a midgut volvulus, lysis of Ladd's bands, placement of the small intestine within the right hemiabdomen and the colon within the left hemiabdomen, with or without a prophylactic appendectomy done to prevent future diagnostic confusion in the setting of acute appendicitis [17]. The Ladd's procedure has been successful in reducing the risk of recurrent volvulus and treating the chronic symptoms associated with intestinal malrotation in adults [1,4,7].

4. Conclusions

Intestinal malrotation in adults is a rare but potentially catastrophic diagnosis. The variability in anatomy creates a range of possible presentations highlighting the need for early consideration of this diagnosis. Our case demonstrates delayed diagnosis in a patient with atypical positioning of Ladd's bands and a chronic/intermittent mid gut volvulus highlighting the importance for the adult surgeon to understand the variability of this diagnosis in adults. Early recognition will facilitate timely management to address the increased morbidity and mortality seen in intestinal malrotation in the adult population.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Sources of funding

No funding was obtained for the development of this paper.

Ethical approval

Not applicable.

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.

Research registration

N/A.

Guarantor

Tiago Ribeiro

Credit authorship contribution statement

TR was responsible for collecting information of the current case, performing a literature review, and writing the first draft of the manuscript. BG was responsible for revision of the manuscript. SB was responsible for revision of the manuscript. HM was responsible for drawing schematic diagrams. PK was responsible for revision of the manuscript and important intellectual content.

All authors read, revised, and approved the final manuscript.

Declaration of competing interest

The authors have no conflict of interest to disclose.

References

[1] S.D. Adams, M.P. Stanton, Malrotation and intestinal atresias, Early Hum. Dev. 5 (2014).
[2] C.A. Hajivassiliou, Intestinal obstruction in neonatal/pediatric surgery, Semin. Pediatr. Surg. 12 (2003) 241–253, https://doi.org/10.1053/j.sempedsurg.2003.08.005.
[3] M.T. Kafadar, A.Y. Cengiz, T. Cavis, I. Bilgic, I. Nadir, Incidental intestinal malrotation in an adult: midgut volvulus, J. Surg. 34 (2018) 337–339, https://doi.org/10.5152/turkjsurg.2017.3468.
[4] J.J. Neville, J. Gallagher, A. Mitra, H. Sheth, Adult presentations of congenital midgut malrotation: a systematic review. [Review], J. Surg. 44 (2020) 1771–1778, https://doi.org/10.1007/s00268-020-05403-7.
[5] P.J. Pickhardt, S. Bhalla, Intestinal malrotation in adolescents and adults: spectrum of clinical and imaging features, J. Roentgenol. 179 (2002) 1429–1435.
[6] D. Nehra, A.M. Goldstein, Intestinal malrotation: varied clinical presentation from infancy through adulthood, Surgery 149 (2011) 386–393, https://doi.org/10.1016/j.surg.2010.07.004.
[7] 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, https://doi.org/10.1016/j.jamcollsurg.2007.11.020.
[8] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, A.J. Beamish, A. Noureldin, A. Rao, B. Vasudevan, B. Challacombe, B. Perakath, B. Kirshtein, B. Ekser, C.S. Pramesh, D.M. Laskin, D. Machado-Aranda, D. Miguel, D. Pagano, F. H. Millham, G. Roy, H. Kadioglu, I.J. Nixon, I. Mukherjee, J.A. McCaul, J. Chi-Yong Ngu, J. Albrecht, J.G. Rivas, K. Raveendran, L. Derbyshire, M.H. Ather, M. A. Thorat, M. Valmasoni, M. Bashashati, M. Chalkoo, N.Z. Teo, N. Raison, O. J. Muensterer, P.J. Bradley, P. Goel, P.S. Pai, R.Y. Afifi, R.D. Rosin, R. Coppola, R. Klappenbach, R. Wynn, R.L. De Wilde, S. Surani, S. Giordano, S. Massarut, S. G. Raja, S. Basu, S.A. Enam, T.G. Manning, T. Cross, V.K.L. Karanth, V. Kasivisvanathan, Z. Mei, S.C.A.R.E. The, Guideline: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) (2020) 226–230, https://doi.org/10.1016/j.ijsu.2020.10.034.
[9] 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. 50 (2018) 46–49, https://doi.org/10.1016/j.ijscr.2018.07.007.
[10] J.H. Soffers, J.P. Hikspoors, H.K. Mekonen, S.E. Koehler, W.H. Lamers, The growth pattern of the human intestine and its mesentery, BMC Dev. Biol. 15 (2015) 31, https://doi.org/10.1186/s12861-015-0081-x.
[11] D.M. Powell, H. Biemann Othersen, C.D. Smith, Malrotation of the intestines in children: the effect of age on presentation and therapy, J. Pediatr. Surg. 24 (1989) 777–780, https://doi.org/10.1016/S0022-3468(89)80535-4.
[12] G.N. Ayane, K. Kadimo, Diagnosis and surgical management of congenital intestinal malrotation presenting with midgut volvulus in an adult: high index of suspicion (case report), Pan Afr. Med. J. 1 (2018) 154, https://doi.org/10.11604/pamj.2018.29.154.13910.
[13] S. Singh, A. Das, A.S. Chawla, S.V. Arya, J. Chaggar, A rare presentation of midgut malrotation as an acute intestinal obstruction in an adult: two case reports and literature review, J. Surg. Case Rep. 4 (2013) 72–75, https://doi.org/10.1016/j.ijscr.2012.10.005.
[14] R.K.N.Allard-Picou Ryan M. Crouse, Ethan A. Talbot, K. Ayana, Intestinal Malrotation in the Adult, - Rosalynn K. Nguyen, Ryan M. Crouse, Ethan A. Talbot, Ayana K. Allard-Picou, 2020, Am. Surg. (2020), https://doi.org/10.1177/0003134820947406. (Accessed 24 October 2020).
[15] B.W. Haak, S.T. Bodewitz, C.F. Kuijper, L.M. de Widt-Levert, Intestinal malrotation and volvulus in adult life, J. Surg. Case Rep. 5 (2014) 259–261, https://doi.org/10.1016/j.ijscr.2014.02.013.
[16] C. Grassi, L. Conti, G. Palmieri, F. Banchini, M.D. Dacco', G.M. Cattaneo, P. Capelli, Ladd's band in the adult, an unusual case of occlusion: case report and review of the literature, J. Surg. Case Rep. 1 (2020) 45–49, https://doi.org/10.1016/j.ijscr.2020.04.046.
[17] J.G. Brungardt, S.C. Liebscher, K.P. Schropp, Malrotation correction in the adult population, World J. Surg. (2020), https://doi.org/10.1007/s00268-020-05790-x.