S Afr J Surg. 2024;62:267-271. Online first
https://doi.org/10.36303/SAJS.00191

S Afr J Surg
ISSN 038-2361

PAEDIATRIC SURGERY

Symptomatic omphalomesenteric duct anomalies in children

G Gerçel,[1,2] Aİ Anadolulu[3,4]

[1] *Department of Paediatric Surgery, Şanlıurfa Training and Research Hospital, Turkey*
[2] *Department of Paediatric Surgery, Sancaktepe Şehit Prof. Dr İlhan Varank Training and Research Hospital, Turkey*
[3] *Department of Paediatric Surgery, Mehmet Akif İnan Training and Research Hospital, Turkey*
[4] *Department of Paediatric Surgery, Faculty of Medicine, Istanbul Medeniyet University, Goztepe Prof. Dr Suleyman Yalcin City Hospital, Turkey*

Corresponding author, email: *goncagercel@gmail.com*

Background: We aimed to present our experience with children with symptomatic omphalomesenteric duct (OMD) anomalies and evaluate the patients' characteristics, treatment, and outcomes.
Methods: Records of children who were operated for symptomatic OMD anomalies in Şanlıurfa Training and Research Hospital between October 2018 and November 2022 were retrospectively analysed.
Results: There were 35 patients with a median age of 31.7 (1 day–17 years) weeks, 29 (82.8%) males and six (17.2%) females. The presenting signs were gastrointestinal tract (GIT) obstruction in 17 (48.6%) patients, acute abdomen in 11 (31.4%), umbilical abnormalities in four (11.4%) and rectal bleeding in three (8.6%). All patients presenting with umbilical abnormalities were newborn. These were OMD fistula to skin (n = 1), OMD fistula to umbilical cord hernia sac (n = 1), OMD cyst in umbilical cord hernia (n = 1) and OMD band adherence to umbilical cord hernia sac (n = 1). Meckel's scan was positive in all three patients with rectal bleeding. Surgical findings in patients other than umbilical abnormalities were diverticulitis (with/without perforation) (n = 14), intussusception due to diverticulum (n = 9) and Meckel's band obstruction (n = 8). At surgery, an ileal resection was performed in 19 cases, wedge resection in 10 cases, resection with stapler in five cases and ileocolonic resection in one patient. On histopathological examination, ectopic gastric mucosa was detected in 11 specimens and both gastric and pancreatic tissue in two. There were only two cases of postoperative complications (incisional hernia, n = 1, postoperative colon perforation due to forced manual reduction of intussusception, n = 1) and all patients survived in good condition.
Conclusion: In the present study, GIT obstruction is the primary symptom in patients with symptomatic OMD anomalies, with umbilical anomalies exclusively found in newborns. Surgery is confirmed as the definitive treatment, with wedge resection and simple diverticulectomy being safe but sometimes insufficient. A significant portion of patients might need more complex segmental bowel resections due to severe complications. With ectopic tissue found in about one-third of cases, managing OMD anomalies presents distinct challenges.
Keywords: omphalomesenteric duct, Meckel's diverticulum, children

Introduction

The omphalomesenteric duct (OMD) is an embryonic structure that connects the yolk sac to the primitive gut and contains vessels that provide nutrition to the developing embryo. Normally, it involutes in utero by the sixth to the ninth week of foetal life and the placenta takes over as the primary source of foetal nutrition[1]. Failure that develops at any point during this process results in a spectrum of anomalies including Meckel's diverticulum (MD), umbilical fistulas, sinus tracts, polyps, cysts, and congenital bands.[1,2]

Proximal part persistence of OMD, known as MD, was the first to be described and named by Johann Friedrich Meckel, a German anatomist, in 1809.[3] MD is the most common remnant of the OMD, accounting for 67– 90% of all cases and is also one of the most common congenital anomalies of the digestive tract in children (2–4%).[4-6]

The diagnosis is difficult in symptomatic cases, and they are usually noticed during surgery. In the majority of the cases, MD is clinically silent. Therefore, it is hard to estimate its exact incidence. Due to the rarity of the OMD anomalies, our information is usually based on small series and case reports.[4,5,7]

Although the majority of OMD anomalies are asymptomatic, they may cause serious complications such as inflammation, perforation, bleeding, intestinal obstruction due to bands, volvulus, and intussusception.[6] The surgical management of OMD anomalies encompasses a spectrum of techniques documented within the medical literature, including wedge or segmental resection for MD and comprehensive umbilical exploration in instances of umbilical anomalies. The variability in surgical approach reflects the diverse clinical presentations and anatomical variations of these anomalies. Postoperative lifelong complication rates associated with these surgical interventions are reported to range between 4% and 34%. Complications can vary according to age and are more common under two years of age.[6,7] This broad range highlights the potential for post-surgical challenges and underscores the importance of

carefully considering the most appropriate surgical strategy for each case.

The present study aimed to gather the experiences of a single institution regarding symptomatic OMD anomalies in paediatric patients. It aims to detail the variations in age and presentation methods, describe the surgical interventions performed, and report the outcomes of histopathological analysis.

Materials and methods

A retrospective analysis of patient files and operative logs was done for all patients who underwent surgery with the diagnosis of symptomatic OMD anomalies in Şanlıurfa Training and Research Hospital between 1 October 2018 and 1 November 2022. Incidental diverticulectomies were not included. We recorded the data of patients on age at the time of diagnosis, sex, clinical presentation, diagnostic methods, intraoperative findings, operative technique, histopathologic results, surgical complications, morbidity, and mortality. The results were subjected to a descriptive analysis.

All procedures performed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1964 and later versions. This study was approved by the Institutional Review Board of Harran University (approval number 2022/24.04) and written informed consent for surgery was obtained from all patients.

Results

During the four-year period evaluated, a total of 6238 surgeries were performed in our paediatric surgery clinics. Among these, 35 (0.56%) children underwent operations due to symptomatic OMD anomalies. There were 29 (82.8%) males and six (17.2%) females. The median age of presentation was 31.7 (1 day–17 years) weeks. Five of the patients were newborns. The prominent indications for surgery were gastrointestinal tract (GIT) obstruction, acute abdomen, umbilical anomalies, and rectal bleeding, respectively.

There were 17 (48.6%) patients aged between 20 days and 15 years presenting with signs of gastrointestinal obstruction. All patients had complaints of cramping abdominal pain and bilious vomiting. In addition, one patient had a previous history of intermittent abdominal pain. During surgery, intussusception with MD as the pathological leading point was detected in nine cases. In five patients, there was obstruction due to meso-diverticular bands compression on bowel wall directly and volvulus around the band was found in three patients. The surgical approach for each patient was determined intraoperatively based on the surgeon's assessment of the clinical findings. Decisions were guided by factors such as palpation of tissue indicating ectopic presence, the location of palpated tissue if present, the anatomical configuration of the diverticulum (wide or narrow base), and the need to define the margins of resection in cases where necrosis was detected. Segmental ileal resection was performed in eight patients, simple diverticulectomy with the linear stapler in five, wedge ileal resection in three, and ileocolonic resection including cecum in one patient due to a large area of necrosis caused by volvulus due to meso-diverticular band.

Acute abdominal findings were observed in 11 (31.4%) patients aged between 3 and 17 years. In one of the patients, there was intra-abdominal free air appearance on a standing straight abdominal X-ray taken before the surgery. A grossly inflamed MD was identified in seven patients and perforated diverticulum in three (Figure 1). In one patient, MD had a gangrenous appearance. It had caused intussusception by inverting into the bowel lumen (Figure 2). Eight of the patients underwent segmentary ileal resection and three of them underwent wedge resection.

There were four (11.4%) patients who presented with umbilical abnormalities. All of the patients were newborns. These were OMD fistula to skin with faecal drainage (n = 1),

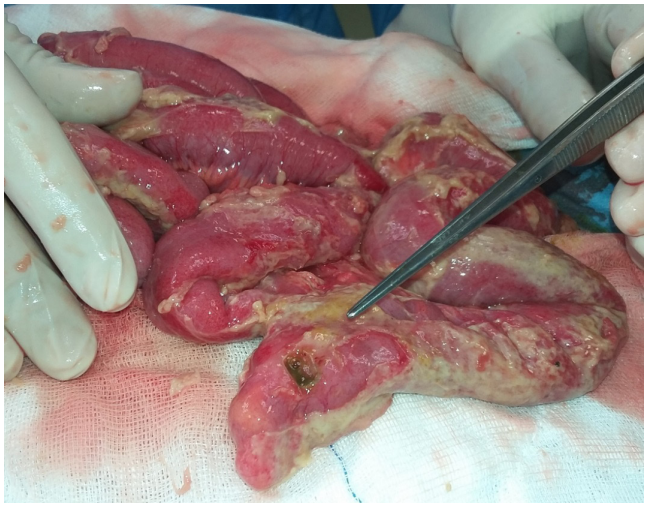

Figure 1: The appearance of perforated diverticulitis at laparotomy

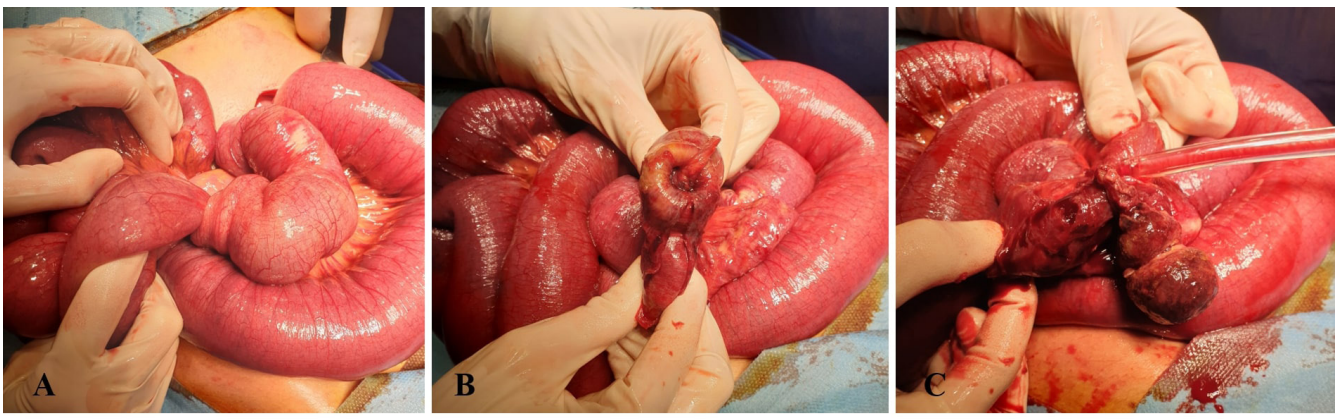

Figure 2: (A) Intussusception due to MD, (B) The appearance of inverted MD, (C) The gangrenous appearance of MD

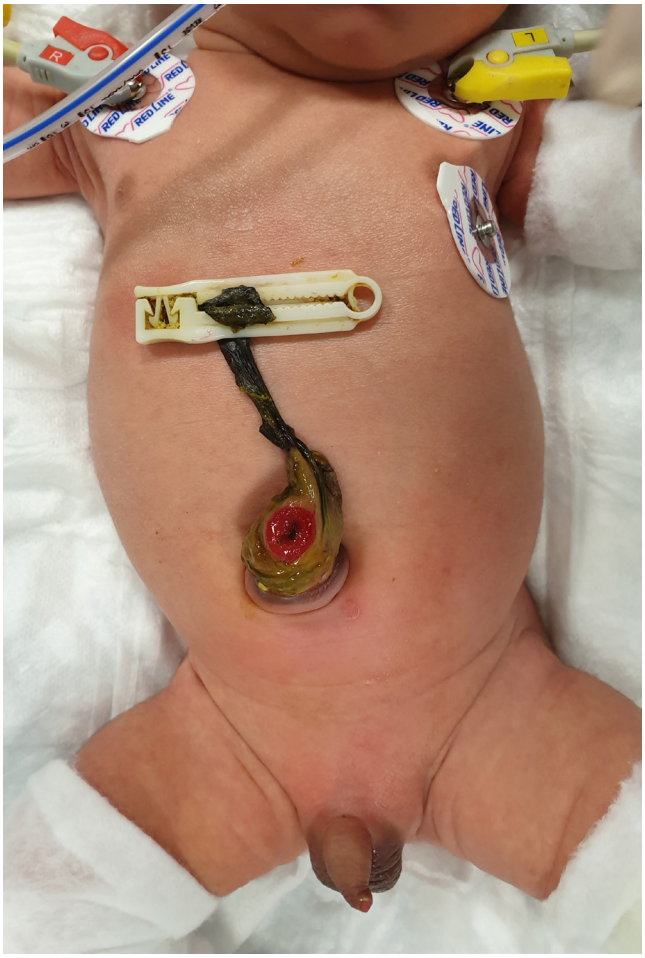

Figure 3: The appearance of OMD fistula to skin

OMD fistula to umbilical cord hernia sac (n = 1), OMD cyst in umbilical cord hernia (n = 1), and OMD band adherence to umbilical cord hernia sac (n = 1) (Figures 3 and 4). Wedge resection was performed in three patients and segmental ileal resection in one.

Painless massive rectal bleeding was the presenting complaint in three (8.6%) patients. They were aged 2, 5 and 10 years. ^{99m}Technetium scintigraphy was performed in all patients and resulted positive in all. Segmental ileal resection was performed in 2 patients and wedge resection in one.

As a total, there were 19 (54.3%) patients who underwent segmental ileal resection followed by end-to-end anastomosis, 10 (28.6%) who underwent wedge resection of ileum, five (14.3%) who underwent simple diverticulum resection with linear stapler and one (2.8%) patient who underwent ileocolonic resection and end-to-end anastomosis.

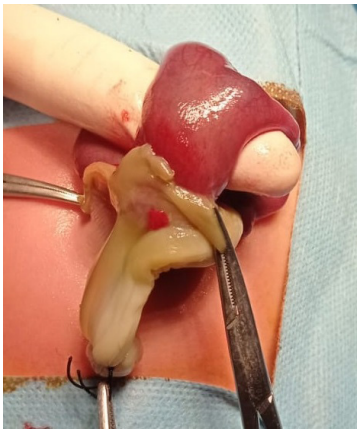

Figure 4: The appearance of OMD fistula to the umbilical cord hernia sac

Postoperative complications developed in two (5.7%) patients, one in the early and one in the late period. One of these patients was a 6-month-old patient who developed intussusception due to MD. During surgery, intussusception was forcefully reduced. A large serosal defect developed in the ascending colon and the defect was primarily repaired. MD was excised with wedge resection. On the postoperative 5th day, the patient's abdominal examination findings worsened and free intra-abdominal air was detected in abdominal X-ray. He was taken to laparotomy again. Perforation was detected in the area where the serosa defect was repaired. An ileocolonic resection anastomosis was performed. Postoperative follow-up was uneventful. The other patient was a 10-month-old patient who had a large area of necrosis caused by volvulus due to meso-diverticular band and underwent ileocolonic resection. Incisional hernia was detected at postoperative 3rd month. Primary repair was performed. There was no additional problem in the follow-up. No mortality was detected in any patient.

Histopathological examination revealed ectopic tissue in 13 (37.1%) specimens. Ectopic gastric mucosa was detected in 11 of them and both gastric and pancreatic tissue were detected in two. Ectopic tissue was detected in four (23.5%) of 17 patients who presented with gastrointestinal obstruction, in six (54.5%) of 11 patients with acute abdomen, in all patients three (100%) with rectal bleeding. The presence of macroscopic appearance suggestive of ectopic tissue was noted during surgery in three of the patients with histopathological detected ectopic tissue. Among patients who had ectopic tissues, nine (69.2%) underwent segmentary intestinal resections, three (23.1%) underwent wedge resections and one (7.7%) simple diverticulum resection with linear stapler at the operation

Table I: Patient characteristics, surgeries performed, and presence of ectopic tissue

Clinical presentation	Number of patients (n, %)	Operation (n), (presence of ectopic tissue, n)				Presence of ectopic tissue (total, n)
		Simple diverticulectomy	Wedge resection	Segmental resection	Ileocolonic resection	
Gastrointestinal obstruction	17 (48.6%)	5 (1)	3	8 (3)	1	4
Acute abdomen	11 (31.4%)	0	3 (2)	8 (4)	0	6
Umbilical anomalies	4 (11.4%)	0	3	1	0	0
Rectal bleeding	3 (8.6%)	0	1 (1)	2 (2)	0	3
Total (n, %)	35 (100%)	5 (14.3%)	10 (28.6)	19 (54.3)	1 (2.8%)	13 (37.1%)

(Table I). In all cases where ectopic tissue was identified, the surgical resection margins were free of ectopic tissue.

Discussion

Although OMD remnants are well-known congenital anomalies, our knowledge is mostly based on small case series and case reports. Its true incidence is difficult to determine as many of them remain asymptomatic.[4,5] While the rate of male and female is reported to be equal in asymptomatic cases, it is reported that it is seen three to four times greater in males in symptomatic cases.[1,8,9] According to a study involving 100 patients with MD, the higher rate of symptomatic cases in men may be due to more acid secretion in the male population as in peptic ulcer disease.[9] There are also publications suggesting that decreased expression of CDX2, a caudal-related homeobox transcription factor protein, is related to increased gastric heterotrophy in MD and downregulation by methylation of this gene is more common in males which may be the other reason.[10] In the present study, it was seen five times greater in males, which is consistent with the literature.

The presentation of OMD anomalies can be quite varied. Symptomatic OMD anomalies vary considerably in clinical scenarios, including intussusception, volvulus, internal hernia, gastrointestinal bleeding, diverticulitis, perforation, umbilical fistulas, sinus tracts and polyps.[1,2] Although, many publications reported that bleeding was the most common presenting symptom in children, some series suggested that obstructive symptoms were prominent. Infectious or inflammatory findings were the least mentioned.[8,9,11,12] In this study, obstructive symptoms were the leading complaints, followed by acute abdominal findings, umbilical anomalies and rectal bleeding, respectively.

Studies have reported that symptomatic OMD anomalies are more common under the age of 10, especially under the age of two years.[7,8] However, it has been reported that obstructive findings are more common under the age of 10 years, while the average age of children presenting with rectal bleeding is higher.[12,13] In this study, 25.7% of the patients were under two years of age and 68.5% were less than 10 years of age. However, the age distribution according to the presenting symptoms was similar between the patients with acute abdominal findings and obstructive symptoms. All umbilical pathologies were predictably younger than the other groups; in this series all were neonates.

It is known that the presence of ectopic tissue increases the possibility of OMD anomalies being symptomatic. Ectopic tissue consists mostly of gastric mucosa but less commonly of pancreatic, jejunal, or colonic tissue. The incidence of ectopic tissue in symptomatic OMD anomalies is reported to be 45–80% in previous series.[8,14,15] In a study of 74 paediatric patients who underwent excision for MD, ectopic tissue was found in 27% of those without symptoms and 74% of the symptomatic patients.[7] In the present study, 37.1% ectopic tissue was detected, which was lower compared to the literature. There was gastric mucosa in 11 patients and gastric and pancreatic tissue association in two patients. Ectopic tissue was found in 54.5% of patients presenting with acute abdomen, 23.5% of patients presenting with obstructive symptoms and all patients presenting with rectal bleeding.

The various degrees of failure during regression of the OMD, present clinically with a wide spectrum of umbilical anomalies ranging from fistulisation of the ileum to the skin, to cysts and polyps.[1,2] As most of these are visible anomalies, they are diagnosed earlier. However umbilical anomalies are relatively rare among OMD anomalies. OMD cyst inside the umbilical cord hernia sac is even rarer and very few cases have been reported in the literature.[16-18] The association of trisomy 13,18 and aneuploidy has been reported in antenatally diagnosed patients.[19,20] In this series, one patient had OMD fistula to skin with faecal drainage and one patient had OMD fistula to umbilical cord hernia sac with mucous drainage. In two patients, the contents of the umbilical cord hernia sac could not be manually reduced to the abdomen before surgery. One of these patients had an OMD cyst in the sac. In the other patient, MD was adherent to the dome of the sac with a meso-diverticular band. No genetic anomaly was detected in any of the patients.

The treatment of symptomatic OMD anomalies is surgical. However, the operative technique of choice is still controversial. Surgery can be performed by wedge resection, segmental intestinal resection or by a simple diverticulectomy. Although publications state that if there is no heterogeneous tissue at the base of the diverticulum with palpation, simple diverticulectomy can be performed, there are also publications advocating that the reliability of palpation alone is not sufficient and the segmental ileal excision should be performed.[12,21,22] In this series, 42.9% of patients underwent excision with methods other than segmental ileal resection. No morbidity was observed in any of the patients who underwent wedge resection and simple diverticulectomy with linear stapler. However, the majority of patients required segmental ileal resection of various lengths due to poor intestinal viability.

Conclusions

In this study, in which symptomatic OMD anomalies were reported, the data indicate that gastrointestinal obstruction was the most common presentation among affected patients. Notably, all instances of umbilical anomalies were identified exclusively in newborns. Surgical treatment has been affirmed as a curative approach for these conditions. While both wedge resection and simple diverticulectomy procedures are established as safe and effective, the study suggests that a considerable number of patients may require more extensive segmental bowel resections, contingent upon the complexity of their clinical presentation and the extent of complications. Furthermore, ectopic tissue was observed in approximately one-third of the symptomatic cases, emphasising the variability and potential challenges in managing OMD anomalies.

Given these findings, it is recommended that surgical teams adopt a flexible approach to the surgical management of OMD anomalies, prepared to escalate from less invasive procedures to more comprehensive resections based on the individual patient's condition and the presence of ectopic tissue. This tailored approach can optimise outcomes, reducing the risk of recurrence and other complications.

Conflict of interest

The authors declare that there is no conflict of interest.

Funding source

No funding was required.

Ethical approval

This study was approved by the Institutional Review Board of Harran University (approval number 2022/24.04). Informed consent was obtained from the patients.

ORCID

G Gerçel https://orcid.org/0000-0003-1395-1764
Aİ Anadolulu https://orcid.org/0000-0002-9742-930X

REFERENCES

1. Sadler TW, Langman J. Langman's medical embryology. 12th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2012. p. 384.
2. Inarejos Clemente EJ, Navarro OM, Navallas Irujo M, et al. Omphalomesenteric duct anomalies in children: A multimodality overview. RadioGraphics. 2021;41(7):2090-110. https://doi.org/10.1148/rg.2021210048.
3. Meckel JF. Über die Divertikel am Darmkanal. Arch Physiol. 1809;9:421-53.
4. Ueberrueck T, Meyer L, Koch A, et al. The significance of Meckel's diverticulum in appendicitis: A retrospective analysis of 233 cases. World J Surg. 2005;29(4):455-8. https://doi.org/10.1007/s00268-004-7615-x.
5. Sancar S, Demirci H, Sayan A, Arıkan A, Candar A. Meckel's diverticulum: Ten years' experience. Turk J Surg. 2015;31(2):65-7. https://doi.org/10.5152/UCD.2015.2834.
6. Durakbasa CU, Okur H, Mutus HM, et al. Symptomatic omphalomesenteric duct remnants in children. Pediatr Surg Int. 2010;52:480-4. https://doi.org/10.1111/j.1442-200X.2009.02980.x.
7. Önen A, Çiğdem MK, Öztürk H, Otcu S, Dokucu AI. When to resect and when not to resect an asymptomatic Meckel›s diverticulum: An ongoing challenge. Pediatr Surg Int. 2003;19:57-61. https://doi.org/10.1007/s00383-002-0850-z.
8. Menezes M, Tareen F, Saeed A, Khan N, Puri P. Symptomatic Meckel's diverticulum in children: A 16-year review. Pediatr Surg Int. 2008;24:575-7. https://doi.org/10.1007/s00383-007-2094-4.
9. Huang CC, Lai MW, Hwang FM, et al. Diverse presentations in pediatric Meckel's diverticulum: A review of 100 cases. Pediatr Neonatol. 2014;55:369-75. https://doi.org/10.1016/j.pedneo.2013.12.005.
10. Martin E, Vanier M, Tavian M, et al. CDX2 in congenital gut gastric-type heteroplasia and intestinal-type Meckel diverticula. Pediatrics. 2010;126:723-7. https://doi.org/10.1542/peds.2009-3512.
11. Hansen CC, Søreide K. Systematic review of epidemiology, presentation, and management of Meckel's diverticulum in the 21st century. Medicine. 2018;97:12154. https://doi.org/10.1097/MD.0000000000012154.
12. Park JJ, Wolff BG, Tollefson MK, Walsh EE, Larson DR. Meckel diverticulum - the Mayo Clinic experience with 1476 patients (1950-2002). Ann Surg. 2005;241:529-33. https://doi.org/10.1097/01.sla.0000154270.14308.5f.
13. Bemelman WA, Huhenholtz E, Heij HA, Wiersma PH. Meckel's diverticulum in Amsterdam: Experience in 136 patients. World J Surg. 1995;19:734-7. https://doi.org/10.1007/BF00295917.
14. Nissen M, Sander V, Rogge P, Alrefai M, Tröbs RB. Meckel's diverticulum in children: A monocentric experience and mini-review of literature. Children. 2022;9(1):35. https://doi.org/10.3390/children9010035.
15. Chen Q, Gao Z, Zhang L, et al. Multifaceted behaviour of Meckel's diverticulum in children. J Pediatr Surg. 2018;53(4):676-81. https://doi.org/10.1016/j.jpedsurg.2017.11.059.
16. Ratan SK, Rattan KN, Kalra R, et al. Omphalomesenteric duct cyst as a content of omphalocele. Indian J Pediatr. 2007;74:500-2. https://doi.org/10.1007/s12098-007-0087-x.
17. Khan YA, Qureshi MA, Akhtar J. Omphalomesenteric duct cyst in an omphalocele: A rare association. Pak J Med Sci. 2013;29(3):866. https://doi.org/10.12669/pjms.293.3581.
18. Mammadov, E. Patent omphalomesenteric duct with protruding bowels through a ruptured omphalocele. Balkan Med J. 2018;35(1):118-9. https://doi.org/10.4274/balkanmedj.2017.0230.
19. Smith GN, Walker M, Johnston S, Ash K. The senographic finding of persistent umbilical cystic masses is associated with lathal aneuploidy and/or congenital anomalies. Prenatal Diag. 1996;16:1141-7. https://doi.org/10.1002/(SICI)1097-0223(199612)16:12<1141::AID-PD2>3.0.CO;2-4.
20. Ross JA, Jurkovic D, Zosmer N, et al. Umbilical cord cysts in early pregnancy. Obstet Gynecol. 1997;89:442-5. https://doi.org/10.1016/S0029-7844(96)00526-1.
21. Pinero A, Martinez-Barba E, Canteras M, et al. Surgical management and complications of Meckel's diverticulum in 90 patients. Eur J Surg. 2002;168(1):8-12. https://doi.org/10.1080/110241502317307508.
22. Williams RS. Management of Meckel's diverticulum. Br J Surg. 1981;68:477-80. https://doi.org/10.1002/bjs.1800680712.