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A Rare Combination of Left-Sided Gastroschisis and Omphalocele in a Full-Term Neonate: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

BCDEF 1 **Tyler Masden**
ABCDEF 2 **Donald C. Moores**
CDEF 3 **Andrei Radulescu**

1 School of Medicine, Loma Linda University, Loma Linda, CA, U.S.A.
2 Division of Pediatric Surgery, Loma Linda University Children's Hospital, Loma Linda, CA, U.S.A.
3 Division of Pediatric Surgery, Loma Linda University Medical Center, Loma Linda, CA, U.S.A.

Corresponding Author: Donald C. Moores, e-mail: DMoores@llu.edu
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Patient: Female, Newborn
Final Diagnosis: Omphalocele
Symptoms: Congenital abdominal wall defect
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Congenital defects/diseases





Background: Left-sided gastroschisis is a rare congenital birth defect characterized by herniation of intra-abdominal organs through an abdominal wall defect to the left of the umbilicus. Approximately half of the 31 cases reported in the literature describe other associated anomalies. To the best of our knowledge, it has never been reported in association with an omphalocele.

Case Report: Here, we present the case of a female newborn, 37 weeks gestational age, born with a 3×6 cm omphalocele and a left-sided gastroschisis with herniation of the small bowel. Both of these anomalies were managed separately, with initial placement of a silo bag on the gastroschisis defect and application of topical agents to the omphalocele until complete epithelialization was achieved. The herniated bowel at the gastroschisis site was reduced with the aid of the silo by 96 hours and the fascia then closed primarily. A gastrostomy tube (G-tube) was placed at 16 weeks of age because of poor oral intake. Definitive closure of the omphalocele and removal of the gastrostomy tube was achieved at 13 months. Her subsequent follow-up visits in the clinic have been uneventful.

Conclusions: Our case report highlights the importance of recognizing this combination of rare conditions and directing appropriate surgical care.

MeSH Keywords: Congenital Abnormalities • Congenital, Hereditary, and Neonatal Diseases and Abnormalities • Gastroschisis • Hernia, Umbilical • Pediatrics

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Background

Gastroschisis is a congenital defect defined by the herniation of intra-abdominal contents through the anterior wall of the abdomen. Left-sided gastroschisis, with the defect to the left of the umbilicus, is far rarer than right-sided gastroschisis. This condition is more associated with other congenital anomalies than its counterpart, with higher occurrence of facial clefting [1].

The underlying pathophysiology behind the development of gastroschisis is still being elucidated. The mainstream model suggests that the right umbilical vein regresses, leaving a weakened fascial region through which the abdominal contents can herniate [2]. A proposed alternative mechanism states that an early disturbance of the omphalo-mesenteric artery leads to the fascial weakness [3]. If either of these theories hold true, further extrapolation implies that disruption of a left-sided omphalo-mesenteric artery or left umbilical vein could result in left-sided gastroschisis [4].

An omphalocele is a congenital midline abdominal wall defect that involves the herniation of intra-abdominal organs into a membrane consisting of internal peritoneum and external amnion. Although the etiology of omphaloceles are unknown, children with omphaloceles have a high prevalence of other congenital malformations (35%) [5]. Small omphaloceles are often managed by primary closure [6]. Large omphaloceles respond well to “delayed external compression reduction of an omphalocele” (DECRO), which involves topical treatment with antibiotics, moistening agents, and compression gauze, with subsequent reduction of the omphalocele to the level of the abdominal wall, followed by fascial closure [6]. Both omphalocele and gastroschisis are often first diagnosed through prenatal sonography [7].

Fortunately, treatment of a left-sided gastroschisis is identical to that of the right-sided form [2]. Primary closure is preferred, but, if not feasible, then a silo bag is used to reduce the small bowel, followed by closure. In this case report we present a unique case of a term female infant with left-sided gastroschisis and an associated omphalocele.

Case Report

We report the case of a female infant born via cesarean section at 37 weeks and 3 days of gestation with a 3×6 cm unruptured omphalocele and a left-sided gastroschisis with an eviscerated small bowel (Figure 1). The prenatal course was significant for maternal history of heroin use during pregnancy and a family history of Beckwith-Wiedemann syndrome. A prenatal ultrasound done 2 weeks prior to birth clearly found an abdominal wall defect suggestive of a large omphalocele

involving the liver, stomach, and bowel. A subsequent prenatal MRI at 36 weeks gestational age was interpreted as a ruptured omphalocele. At the time of delivery, the child was noted to have an intact omphalocele and bowel eviscerated through a separate defect in the left abdominal wall. Apgar scores were 6 and 8 at 1 minute and 5 minutes, respectively. Due to initial respiratory distress the infant was placed on continuous positive airway pressure (CPAP) 5 with 40% FiO₂. The herniated bowel was noted to appear dusky, purple, and edematous at first, but the color rapidly improved upon normalization of the oxygen saturation and repositioning of the bowel. At this point, the decision was made to place the bowel in a Silastic Silo (Bentec Medical) with a plan to reduce it gradually to accommodate the lack of domain in the abdominal cavity (Figure 1). The omphalocele was addressed nonoperatively by treatment with topical antibiotic ointment, followed by Xeroform gauze and Kerlix.

By the fourth day of life, the contents of the silo were reduced to the abdominal wall level and the decision was made to take the patient to the operating room (OR) to close the defect. Adhesions between bowel loops and the chest wall were separated gently, followed by reduction of the hernia and primary closure of the fascia. Of note, the left diaphragm was intact. After irrigation of the wound, the skin was close. (Figure 1).

By 16 weeks, the omphalocele had responded well to topical treatment and was completely epithelialized, and the gastroschisis site had completely healed. However, the child had problems tolerating full feeds, so open gastrostomy tube placement was performed.

At 13 months, the infant had good progression in weight and development, the contents of the omphalocele were reducible, and the child was eating well, so a decision was made to close both the omphalocele defect and the gastrostomy. The omphalocele defect was able to be closed primarily without mesh. A pseudo-umbilicus was successfully recreated in the midline (Figure 2). The postoperative course was uneventful and the patient was seen at follow-up clinic visits over the past year.

Discussion

First described by Blair et al. in 1988, left-sided gastroschisis is an unusual congenital malformation that has only been reported 31 times in the literature, making it exceptionally rare [8]. What makes the present case unique is the presence of an associated omphalocele. It is very common for omphaloceles to be associated with other congenital malformations, with associated defects being present as often as 35% of the time [5]. Gastroschisis has a mortality rate of 4%, whereas omphaloceles

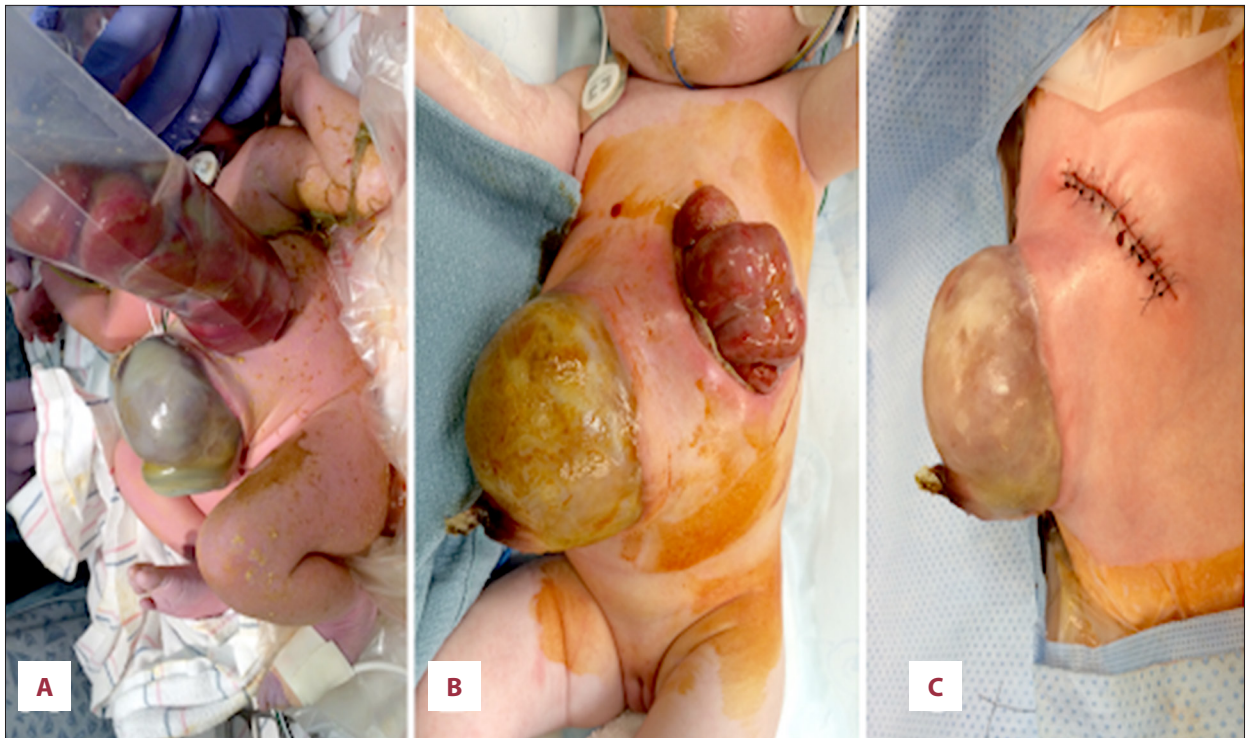


Figure 1. (A) Omphalocele and a left-sided gastroschisis containing eviscerated small bowel in a silo; (B) Following removal of the silo before closure; (C) After closure of left-sided gastroschisis.



Figure 2. Before (A) and after repair of the omphalocele and takedown of the gastrocutaneous fistula (B).

has a higher mortality rate of 18%, which is attributed to the presence of associated congenital malformations [5].

Although congenital deformities can occur at any point during gestation, the first trimester is considered the most vulnerable time period. Some malformations have been linked to the presence of risk factors, called teratogens. The etiology behind omphaloceles, however, is still a mystery. Omphaloceles are not usually associated with exposure to teratogens, although it has been shown that mothers of infants with omphaloceles have higher consumption of alcohol and were more commonly reported to be heavy smokers [9].

Gastroschisis has also been shown to be increased in women who smoke and have higher use of alcohol [9]. A case-controlled study funded by the CDC found a 2-fold increase in the risk of gastroschisis and other congenital malformations when the birth mother was using opioids during the first trimester of pregnancy [10]. In the present case, the mother's first-trimester use of heroin may have had a teratogenic effect and led to the subsequent congenital malformations.

Prenatal ultrasound for gastroschisis and omphalocele is independently of high value, but we believe that for more complex abdominal wall defects, such as this condition, prenatal MRI could prove to be of even greater value. In this case, the MRI was understandably misinterpreted due to the fact that this condition has never previously been reported.

The underlying cause of left-sided gastroschisis is unknown. Previously written and discussed theories have been unable to explain why the vast majority of cases occur to the right of the umbilicus. One suggested model states that early regression of the left umbilical vein may lead to left-sided gastroschisis. Other models suggest that the true origin lies in an abnormality of morphogenesis, with a right-left axis switch initiating the presence of the abnormality on the opposite side [11,12].

Suver et al. [13], in a case report series, showed that neonates with left-sided gastroschisis have different associations than those with right-sided defects. Left-sided defects were more frequent in females and had higher ratios of extraintestinal

anomalies. These unique features may indicate the presence of a genetic, hormonal, or embryological cause for left-sided defects compared with their more common counterparts [14].

Some authors believe that omphalocele and gastroschisis have a common origin, with gastroschisis representing an in-utero rupture of an omphalocele. However, this theory does not explain the accepted epidemiologic and genetic differences between gastroschisis and omphalocele [14].

Another proposed theory that could partially explain our patient's double abdominal wall defect describes gastroschisis as originating from the improper behavior of abdominal folds [12]. These folds normally allow for abdominal closure, influenced by the yolk stalk originating slightly to the right side of the body. As the yolk stalk merges with the connecting stalk in an area to the right of midline, the wall fold fails to close, leading to the possible presence of gastroschisis. If this were to occur on the opposite side of the body, it could lead to a left-sided gastroschisis [15].

The occurrence of a left-sided gastroschisis is much rarer than a right-sided defect and should alert one to the possible presence of associated congenital anomalies.

Conclusions

Left-sided gastroschisis, in and of itself, is a very rare congenital anomaly. To the best of our knowledge, left-sided gastroschisis with an associated omphalocele has never been previously reported. Prenatal ultrasound can be extremely helpful in delineating and monitoring abdominal wall defects and other anomalies, and prenatal MRI can provide even greater anatomic detail. This report shows that these 2 conditions are not mutually exclusive. Although this is a rare association, the surgical management of this condition can be addressed with standard surgical techniques.

Conflict of interest

None.

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