Abdominal Wall Defects

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Practice Gaps

Abdominal wall defects are a relatively common congenital anomaly encountered in the pediatric population. These defects include 2 separate pathologies, gastroschisis and omphalocele, with divergent pathophysiologic origins, clinical manifestations, and management strategies. Although the mode and timing of delivery is somewhat controversial, particularly for gastroschisis, most of the evidence supports delivery at a high-volume tertiary care center with immediate access to neonatal and pediatric surgical expertise. Clinicians should be aware of a rare variant of gastroschisis, closing gastroschisis, because early recognition and treatment may affect patient outcomes, as well as complicated gastroschisis and giant omphalocele because of the more challenging surgical considerations.

Abstract

The 2 most common congenital abdominal wall defects are gastroschisis and omphalocele. Both are usually diagnosed prenatally with fetal ultrasonography, and affected patients are treated at a center with access to high-risk obstetric services, neonatology, and pediatric surgery. The main distinguishing features between the 2 are that gastroschisis has no sac and the defect is to the right of the umbilicus, whereas an omphalocele typically has a sac and the defect is at the umbilicus. In addition, patients with an omphalocele have a high prevalence of associated anomalies, whereas those with gastroschisis have a higher likelihood of abnormalities related to the gastrointestinal tract, with the most common being intestinal atresia. As such, the prognosis in patients with omphalocele is primarily affected by the severity and number of other anomalies and the prognosis for gastroschisis is correlated with the amount and function of the bowel. Because of these distinctions, these defects have different management strategies and outcomes. The goal of surgical treatment for both conditions consists of reduction of the abdominal viscera and closure of the abdominal wall defect; primary closure or a variety of staged approaches can be used without injury to the intra-abdominal contents through direct injury or increased intra-abdominal pressure, or abdominal compartment syndrome. Overall, the long-term outcome is generally good. The ability to stratify patients, particularly those with gastroschisis, based on risk factors for higher morbidity would potentially improve counseling and outcomes.

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Objectives

After completing this article, readers should be able to:

1. Distinguish between gastroschisis and omphalocele.
2. Identify the major prenatal ultrasound findings of the congenital abdominal wall defects.
3. Recognize the rare variant of gastroschisis, closing gastroschisis.
4. Describe the management and surgical techniques used for patients with gastroschisis and omphalocele, including giant omphalocele.
5. Recognize the clinical manifestations of abdominal compartment syndrome and its treatment.

INTRODUCTION

The 2 most common congenital abdominal wall defects are gastroschisis (Fig 1A) and omphalocele (Fig 1B). Both are typically diagnosed prenatally using fetal ultrasonography, and affected patients are treated at a center with access to high-risk obstetric services, neonatology, and pediatric surgery. In this review, we discuss the distinguishing features, current management strategies, and outcomes of patients with these defects.

EPIDEMIOLOGY AND PATHOPHYSIOLOGY

The incidence of congenital abdominal wall defects has been increasing, primarily because of the increased incidence of gastroschisis. (1) Gastroschisis occurs in 1 in approximately 4,000 live births (2) with a male preponderance and has become the most common abdominal wall defect over the past 30 years. (3) A strong association with young maternal age has been noted. The overall incidence of omphalocele is 1 to 2.5 per 5,000 live births. (4)

An omphalocele results from the failure of the bowel loops to return to the abdominal cavity after the physiologic herniation through the umbilical cord that occurs between the 6th and 11th week of development. Several mechanisms have been proposed for the pathogenesis of gastroschisis. One theory is that the defect arises from failure of the umbilical coelom to develop, leading to rupture of the elongating intestine out of the body wall to the right of the umbilicus. An alternative explanation is that the embryonic structures fail to incorporate into the umbilical cord. In addition, experts suggest that several environmental exposures and demographic risk factors contribute to its development.

GASTROSCHISIS

Clinical Aspects

Gastroschisis is usually less than 4 cm in diameter, has no covering membrane or sac, and generally contains only small intestine, potentially with the stomach or gonad. In almost all cases, it is present to the right of the umbilical cord. (5) After birth, the bowel may appear fairly normal or may be thickened, matted, and covered with a fibrinous peel. In contrast to patients with omphaloceles, those with gastroschisis do not typically have associated congenital anomalies but are more likely to have abnormalities of the bowel, including atresias. Many affected patients are born preterm and are often small for gestational age. Those with atresia, perforation, necrosis, or volvulus fall into a separate category called "complicated gastroschisis."

Gastroschisis is commonly seen on mid-second trimester fetal ultrasonography with characteristics of a right-sided defect with free-floating bowel in the amniotic cavity. There are a few ultrasonographic findings that raise the concern for intestinal complications; of these intra-abdominal bowel dilation appears to be the most reliable predictor of complex gastroschisis. (6) In addition, elevated α-fetoprotein concentrations in both maternal blood and amniotic fluid have been correlated with gastroschisis.

Closing or closed gastroschisis is a rare variant of complicated gastroschisis in which the defect narrows in utero, resulting in strangulation and subsequent ischemia of the herniated bowel and atresia. The most severe cases can lead to complete loss of the midgut with short gut syndrome (Fig 2) The patient depicted was diagnosed with closing gastroschisis and initially had complete atresia and significant bowel loss (Fig 2A and 2B). After exploration 6 weeks later, the bowel had grown with significant progressive

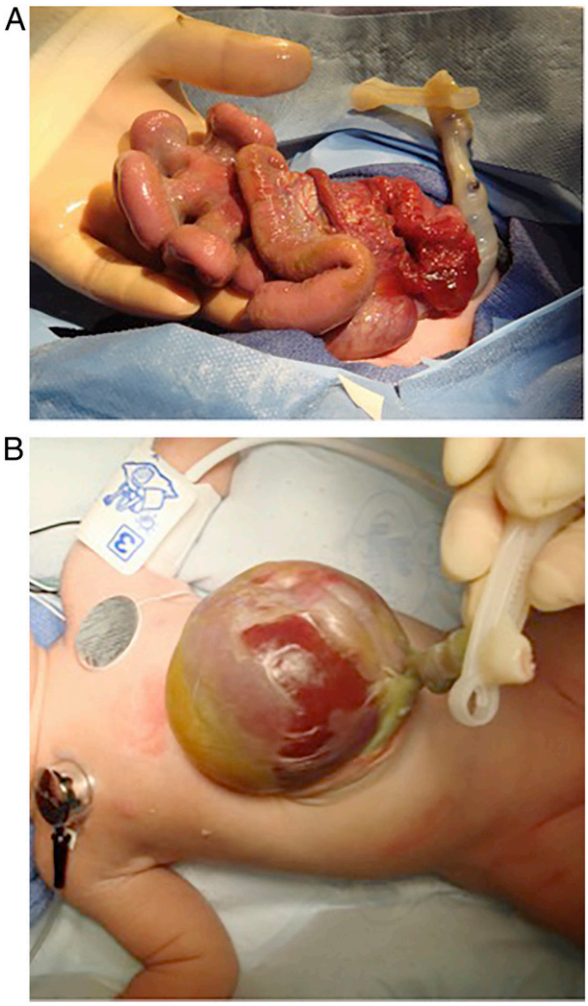

Figure 1. A. Picture of gastroschisis with no sac and the defect to the right of the umbilicus. B. Picture of omphalocele with a sac present and defect at the umbilicus.

development of bowel loops (Fig 2C) and anastomosis and complete closure was achieved. This patient was discharged 9 days later on full feedings. This is one of a series of 5 infants treated at a single referral center for closing gastroschisis (A.P., personal communication, 2016). The defects found in these patients ranged from 0.5 to 2 cm and all tolerated oral intake at discharge, with 3 requiring supplemental nutrition. Affected patients have variable outcomes that depend on the amount of bowel that is viable but resulting in significantly higher morbidity, mortality, and short bowel syndrome rates. If suspected on prenatal imaging, preterm delivery may be indicated.

Management

The optimal mode and timing of delivery for patients with gastroschisis are controversial. Some experts have advocated the use of routine cesarean delivery to avoid injury to the

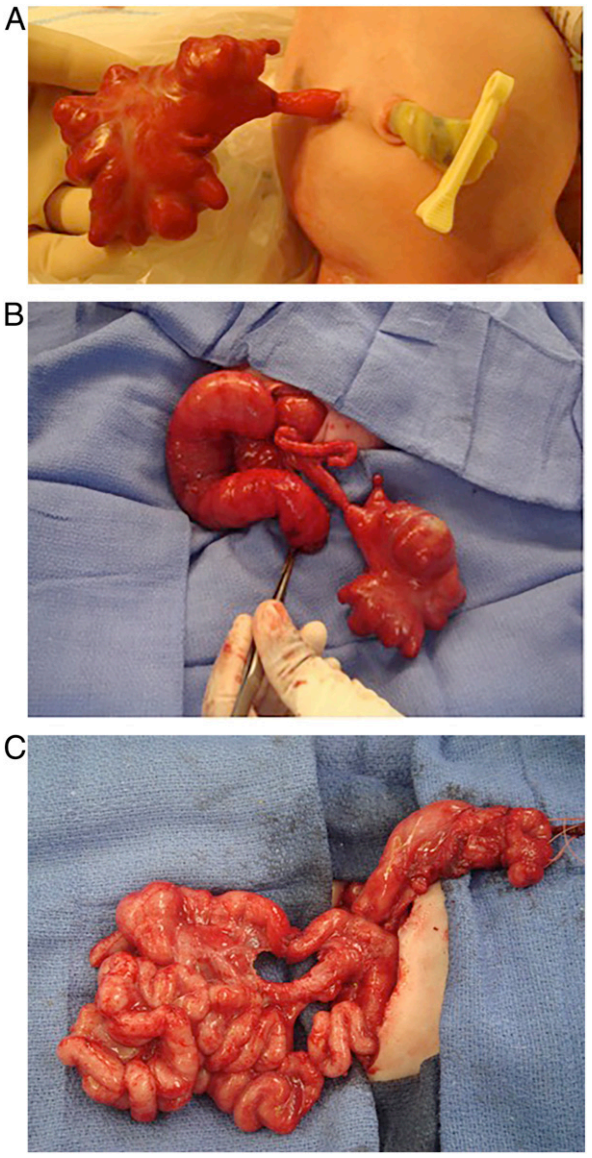

Figure 2. A. Picture of closing gastroschisis with a single strand of tissue (superior mesenteric artery) and complete separation of the midgut (atresia). B. Intraoperative exploration at birth, of the same patient as shown in A, demonstrating complete atresia and bowel loss. C. Same patient 6 weeks later, demonstrating growth of small bowel left in situ.

exposed bowel, but published literature has not shown a difference in outcomes in infants delivered via cesarean versus vaginal delivery. (7) Similarly, certain centers perform early delivery of the fetus to reduce the inflammatory peel on the bowel. However, data have not shown conclusive evidence supporting this view, and the risks associated with prematurity argue against this practice. (8)(9) Thus, the delivery method should be at the discretion of the obstetrician and parents. Most authors and clinicians encourage delivery at a tertiary center with immediate neonatal and pediatric surgery access. (10)(11) The Canadian Pediatric

Surgery Network reviewed data on infants with gastroschisis from 18 pediatric surgical centers and concluded that delivery outside a perinatal center requiring transfer was a significant predictor of complications. (12)

Once the infant is born, fluid resuscitation and gastric decompression should be initiated immediately. Given the significant evaporative and heat loss these patients experience because of the exposed viscera, the bowel must be wrapped in warm, saline-soaked gauze and the lower half of the infant placed in a bowel bag.

The primary goal of surgical repair is to place the intestine back into the abdominal cavity without trauma to the bowel or to avoid increased intra-abdominal pressure. The bowel should be inspected for obstructing bands, matting, perforation, or atresia. Various options for surgical treatment are available, including:

- Primary reduction with surgical fascial closure
- Silo placement with serial reductions and delayed surgical closure of the fascia
- Primary reduction without fascial closure
- Delayed reduction without fascial closure

The last 2 surgical procedures are commonly referred to as "sutureless" closure.

Primary reduction in the operating room involves transport to the operating room, general anesthesia, division of the umbilical vessels and urachus, and suturing of the fascia and skin. Alternatively, surgeons may place a spring-loaded preformed silo into the abdominal defect at the bedside (Fig 3). (13) Serial reductions are then performed daily or twice a day with the aid of gravity until the contents have reached the level of the fascia. This slow reduction allows the bowel edema to be gradually reduced and allows for bowel reduction without increasing intra-abdominal pressure. It is important that the reduction be performed over 3 to 5 days. Any type of surgical closure or sutureless closure can then be done.

Sutureless closure entails covering the abdominal defect with the umbilical cord or synthetic dressing such as a self-adherent foam dressing and allowing closure by secondary intention. We have reported a technique of primary sutureless closure of gastroschisis using negative pressure dressing/wound vacuum (14) (Fig 4). This procedure involves initial placement of a silo with gradual reduction of the intra-abdominal contents. Subsequently, the defect is primarily closed with adhesive tape and wound vacuum. This procedure can be performed at the bedside without anesthesia and without going to the operating room. It has the advantage of gentle silo reduction without increasing the intra-abdominal pressure and causing compartment syndrome. It is also an easily reversible procedure because the adhesive tape and the wound vacuum can be easily removed if the abdominal pressure rises after closure. A randomized

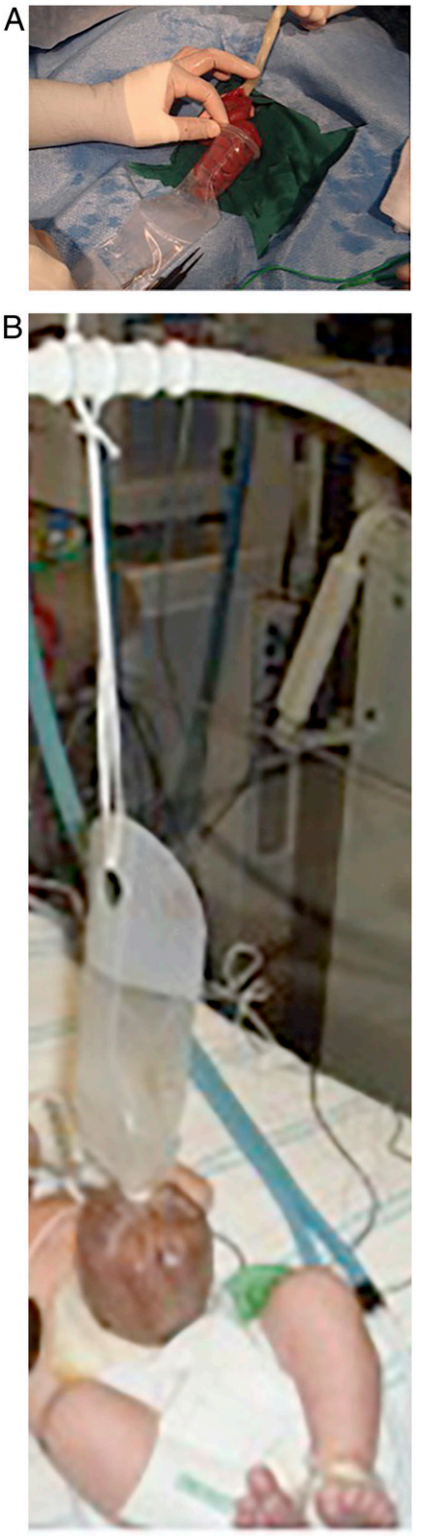

Figure 3. A. Intraoperative picture of silo being placed. B. Silo placed and held upright for gravity to aid with reduction of bowel.

control study comparing sutureless versus sutured gastroschisis closure found no difference in complications. (15) Advantages of this method include the lack of need for transport, potential avoidance of anesthesia, and improved cosmetic result. Most series report a hernia rate of 60% to 84%, of which most close spontaneously; with the wound vacuum closure, the hernia rate is much lower. (16) Nonabsorbable mesh or biosynthetic patches can also be used for closure when primary fascial closure cannot be achieved.

Abdominal compartment syndrome can be a complication after reduction of the bowel. Intra-abdominal pressures greater than 15 to 20 mm Hg indicate compartment syndrome. This pressure can be determined with the use of intragastric or intravesical catheters. Concerning signs also include increased peak or mean inspiratory pressures, need for vasopressor support, or metabolic acidosis. Immediate decompressive laparotomy or release of the closure with silo placement should be undertaken if abdominal compartment syndrome is suspected. Given this complication, the approach for the type of closure must be decided based on conditions such as prematurity, abdominal domain, and degree of respiratory distress.

For patients with gastroschisis and an associated atresia or perforation, the management is more complex. Care of these infants must be individualized based on their gestational age, weight, and clinical status as well as the length and condition of the bowel. The possible techniques include primary anastomosis with closure if the bowel is in good condition; creation of stomas with closure; or reduction of unrepaired bowel into the abdomen with closure and repeat surgery for establishment of bowel continuity in the future.

Postoperatively, it is common to have delayed recovery of bowel function as a result of abnormal intestinal motility, which is frequently observed in these patients. During this period of dysmotility, gastric decompression and parenteral nutrition should be provided until enteral feedings are

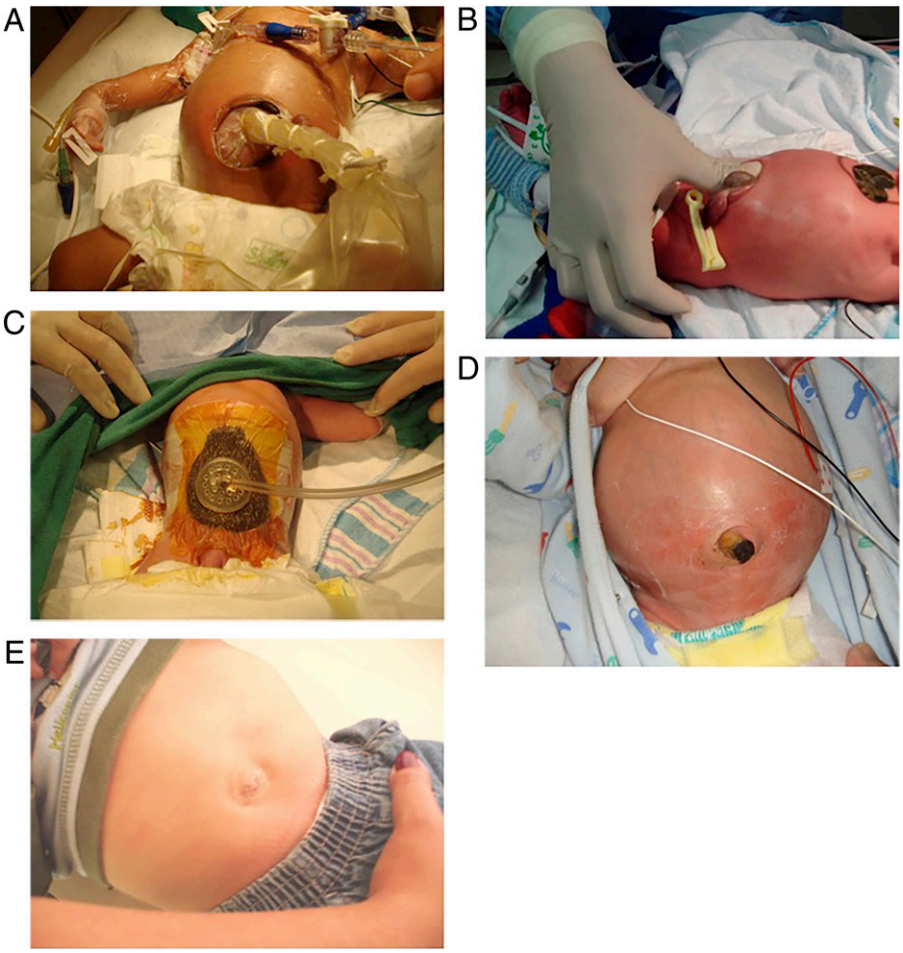

Figure 4. A. Picture demonstrating serial reductions with umbilical ties until at the level of the fascia. B. Complete reduction of gastroschisis. C. Adhesive tape and wound vacuum applied to gastroschisis. D. Picture after wound vacuum removed, demonstrating closure. E. Picture of abdomen 6 weeks after surgery.

started. If bowel improvement is not observed after 4 to 6 weeks, imaging studies can be performed to evaluate for the presence of an intestinal atresia which is often difficult to visualize because of matted bowel.

Prognosis

Long-term outcomes and survival of patients with gastroschisis are generally excellent, with survival rates greater than 90% in large series. (17)(18) Outcomes are poorer in patients with an associated finding such as atresia, perforation, necrosis, or volvulus. (19) However, a single-center study focusing on quality of life using a validated survey demonstrated high average quality of life scores that were independent of severity, after the age of 2 years, which were comparable to published outcomes of healthy children. (20) Potential long-term issues that can be seen in these patients include cholestasis, recurrent, nonspecific abdominal pain, bowel obstruction, and need for scar revision.

OMPHALOCELE

Clinical Aspects

Omphalocele is a large defect, usually greater than 4 cm, covered by an amniotic membrane, which contains intestines and other abdominal organs including the liver and often the spleen and gonad. (5) Patients with an omphalocele often have other congenital anomalies, chromosomal abnormalities, or syndromes. In addition, omphaloceles can be combined with pentalogy of Cantrell, cloacal exstrophy, and the rare omphalocele, exstrophy of the bladder, imperforate anus, and spinal anomaly (OEIS) complex.

Infants with an omphalocele are typically diagnosed prenatally. The fetal ultrasound characteristics include a contained herniation in a membranous sac. Additional associated anomalies may also be identified on prenatal ultrasonography; however, up to one-third of patients with isolated defects are found to have other abnormalities postnatally.

A giant omphalocele contains liver and has a defect of at least 5 to 10 cm in diameter. In addition to an underdeveloped abdominal wall cavity, these patients commonly have pulmonary hypoplasia as well. Giant omphaloceles are associated with a high morbidity and mortality rate. The operative treatment for these patients is also challenging. (21)

Management

Most patients with an omphalocele are born at term gestational age. Some experts advocate for cesarean delivery if there is an extra-abdominal liver to avoid hepatic injury during a vaginal delivery. However, neither type of delivery has been shown to be superior.

Initial management involves obtaining intravenous access and initiating fluid resuscitation as well as gastric decompression with a naso- or orogastric tube. An assessment of the neonate's cardiopulmonary system and complete evaluation for associated anomalies is mandatory. As such, an echocardiography and abdominal ultrasonography should be performed. In addition, a blood glucose level should be checked because hypoglycemia may be an indication of Beckwith-Wiedemann syndrome, which occurs in 12% of patients with an omphalocele.

The management approach for infants with an omphalocele depends on the defect size, birth gestational age and

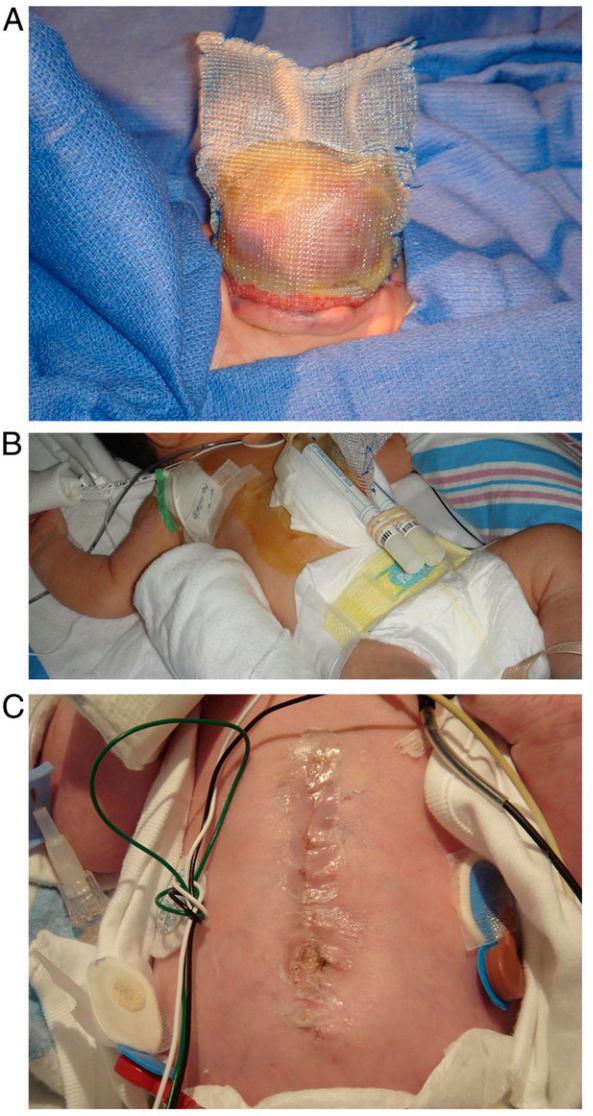

Figure 5. A. Primary closure of a large omphalocele with placement of mesh. B. Reduction of contents. C. Complete closure in 5 days.

weight, and the existence of associated anomalies. In a stable patient with a small defect, primary repair with surgical closure may be possible. The sac may be removed or inverted before fascial closure. If the sac is adherent to the liver, some defects may need to be left in place to avoid liver injury and hemorrhage. However, more commonly, because of the size of the defect, loss of domain of the peritoneal cavity, or instability of the infant, primary closure is not possible, and various techniques are used for coverage and closure. Staged or delayed closure (Fig 5 and Fig 6) of the defect is typically used. Eschoratic therapy, sometimes referred to as the "paint and wait" technique, is often used, in which a topical agent, most commonly silver sulfadiazine, is applied to the sac daily. It creates a gradual eschar with subsequent epithelialization, leaving a ventral hernia. This process takes weeks to months to complete and may be combined with compressive dressings once the sac is thick enough to slowly reduce the contents into the abdomen. Later closure may involve mobilization of skin flaps, component separation, (22) use of tissue expanders, (23) or a patch. (24) A recent report describes a series of patients using a serial taping method to gradually reduce the abdominal contents. (25) With all of these techniques, it is important to avoid kinking of the hepatic veins that may occur with reduction of the liver. This can lead to a metabolic acidosis and may require reoperation to reorient the position of the liver. Another potential complication that can arise either before primary repair or while undergoing topical therapy before the eschar has completely formed is rupture of the sac. A range of methods can be used to manage a ruptured sac, depending on the size of the tear and status of the infant, and includes suture repair, skin closure, and placement of a patch.

Prognosis

The main determinant of prognosis for infants with an omphalocele is the association with structural or chromosomal anomalies that may occur in as many as 80% of affected patients. Major cardiac anomalies are seen in approximately one-third of patients with omphaloceles. Survival rates range from 70% to 95%, with most of the mortality arising from associated anomalies. (26)(27) In addition, a number of long-term medical problems have been found in patients with large omphaloceles, including gastroesophageal reflux disease, pulmonary insufficiency, asthma, and feeding difficulties. (28)(29) Patients with giant omphaloceles have increased morbidity because of an increased visceroabdominal disproportion leading to prolonged mechanical ventilation and a longer hospital stay.

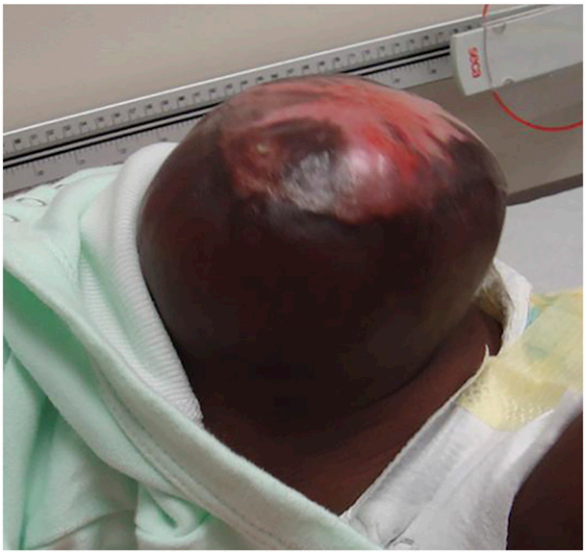

Figure 6. Delayed closure technique used for a large omphalocele after painting with silver sulfadiazine demonstrating complete epilethialization after 8 months.

FUTURE DIRECTIONS

Future goals in the care of patients with gastroschisis are mostly directed toward preventing damage to the exposed bowel as a result of amniotic fluid. Amniotic fluid exchange, (30)(31)(32) nitric oxide, (33) diuretics, (34) and fetoscopic surgery (35)(36)(37) have been tried in animal models with limited success. The other areas of focus include the timing of delivery and the role of intra-abdominal bowel dilation, as discussed herein.

American Board of Pediatrics Neonatal-Perinatal Content Specification

- Know the morphogenesis of the GI tract and factors that lead to congenital malformations.

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1. A woman presents for antenatal ultrasonography at 20 weeks' gestation. The fetus is noted to have probable gastroschisis. Which of the following statements concerning gastroschisis is correct?
    A. It is more common in girls.
    B. It usually presents with an intact peritoneal sac at this gestational age.
    C. The defect typically occurs to the left of the umbilicus.
    D. The incidence is approximately 1 in 4,000 live births and has become the most common abdominal wall defect.
    E. In the current era, it is almost always associated with either marijuana or heavy tobacco use during the first trimester.
2. A female neonate with gastroschisis is born after the mother has preterm labor at 29 weeks' gestation. Which of the following is most likely to be seen in a neonate with gastroschisis?
    A. Large for gestational age.
    B. Congenital anomalies of the heart and brain.
    C. A defect that is almost always larger than 10 cm.
    D. Abnormalities of the bowel such as an atresia.
    E. Vocal cord paralysis.
3. Your team is planning the delivery and postdelivery care for a patient with gastroschisis. Which of the following practices is an appropriate component of the routine care for gastroschisis?
    A. Cesarean delivery, regardless of labor status.
    B. Delivery before 32 weeks of gestation.
    C. Avoidance of nasogastric tube insertion.
    D. Immediate nasal continuous positive airway pressure administration.
    E. Placement of the bowel in warm, saline-soaked gauze and the lower half of the body in a bowel bag.
4. A woman undergoes antenatal ultrasound evaluation and the fetus is noted to have omphalocele. Which of the following statements concerning omphalocele is correct?
    A. The defect is usually greater than 4 cm and covered by an amniotic membrane.
    B. It is almost always an isolated defect, with no other anomalies present.
    C. A "giant" omphalocele refers to the condition in which the entire intestinal tract is located outside the body.
    D. It is associated with high rates of very preterm birth.
    E. There is definitive evidence that cesarean delivery improves outcomes for both the mother and neonate.
5. An infant born at term gestational age has been treated in the NICU for gastroschisis for several weeks. The patient has been able to work up to full enteral feedings. Arrangements are being made for transition to the home. Which of the following is the main determinant of prognosis?
    A. Race/ethnicity.
    B. Presence of structural or chromosomal anomaly.
    C. Presence or absence of intraventricular hemorrhage.
    D. Sex.
    E. Receipt of antenatal or postnatal steroids.

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This journal-based CME activity is available through Dec. 31, 2022, however, credit will be recorded in the year in which the learner completes the quiz.

2020 *NeoReviews* is approved for a total of 10 Maintenance of Certification (MOC) Part 2 credits by the American Board of Pediatrics (ABP) through the AAP MOC Portfolio Program. *NeoReviews* subscribers can claim up to 10 ABP MOC Part 2 points upon passing 10 quizzes (and claiming full credit for each quiz) per year. Subscribers can start claiming MOC credits as early as May 2020. To learn how to claim MOC points, go to: https://www.aappublications.org/content/moc-credit.

Abdominal Wall Defects

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