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Abstract

Gastroschisis is a congenital anterior abdominal wall defect which results in herniation of intra-abdominal contents early in utero. Prenatal ultrasonography has become the optimum means of diagnosing gastroschisis. Knowing the diagnosis in advance allows for appropriate resources to be available to facilitate the delivery at or near a tertiary neonatal care center. Meticulous perioperative management is imperative for good patient outcomes. Abdominal closure can be performed primarily or using a staged technique.

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Congenital or acquired complications, e.g., atresias, perforation, and delayed necrotizing enterocolitis, must be identified promptly and managed carefully. Patient outcomes in gastroschisis are typically excellent, especially if close attention is paid to the details of perioperative management and surgical technique.

Keywords

Gastroschisis · Abdominal wall defects · Intestinal atresias · Silo · Prenatal diagnosis

Introduction

Gastroschisis is an anterior abdominal wall defect that occurs early in fetal development, which results in herniation of intra-abdominal viscera into the amniotic sac. This typically occurs to the right of the umbilical stalk (Fig. 1). The prevailing hypothesis is that the defect occurs at the site of involution of the second (right) umbilical vein. Because most or all of the midgut is outside of the peritoneal cavity, this anomaly is accompanied by nonrotation of the bowel and an increased incidence of other intestinal abnormalities, including atresia, perforation, and infarction, resulting from midgut volvulus or vascular thrombosis. Gastroschisis is much more common now than omphalocele with an incidence of approximately 2.5 per 10,000 live births. Most infants with gastroschisis are born prematurely (35–37 weeks'

gestation), weighing 2000–2500 g. The defect is almost always to the right of the umbilicus and generally measures 2–3 cm in diameter. In addition to the stomach and urinary bladder, the transverse and left colon may be extracoelomic. Also visible may be the testicles in males and the fallopian tubes and ovaries in females. The intestine is foreshortened and edematous and generally has a fibrin coating (Fig. 2). Atresia involving the small or large intestine occurs more often (10–20%) than in patients with omphalocele (~1%). The most striking difference in appearance between omphalocele and gastroschisis is the absence of a sac or membrane covering the herniated contents in gastroschisis (Christison-Lagay et al. 2011).

History

The first successful surgical repair of gastroschisis was performed by Watkins in 1943. Although there were improvements in the perioperative management and surgical procedures for gastroschisis, the mortality remained significant and was reported to be as high as 90%. Two major advances occurred in the late 1960s that led to a dramatic improvement in the



Fig. 1 An infant with gastroschisis. Note herniated contents to the right of the umbilical stalk



Fig. 2 Foreshortened, edematous bowel

survival of infants with gastroschisis. In 1967, Schuster et al. described a technique of staged reduction of the herniated bowel and abdominal closure for patients in whom primary closure was not possible. This allowed for more rapid bowel recovery and decreased risk from sepsis. The second major advance was the evolution of intravenous nutrition, which, remarkably, allowed for growth and development during the prolonged period that these infants could not tolerate enteral feeding. Over the past four decades, the outcome for infants with gastroschisis has dramatically improved, resulting in a survival rate that is now greater than 90% (Holland et al. 2010).

Prenatal Considerations

Fetal ultrasonography can detect gastroschisis as early as the first trimester of pregnancy. Sonographic detection of extracoelomic bowel with no covering membrane strongly suggests the diagnosis. Other findings such as bowel dilatation and/or bowel wall thickening/edema are concerning for bowel obstruction and/or ischemia (Kuleva et al. 2012; Long et al. 2011). However, the degree of bowel dilatation is not necessarily indicative of the extent of intestinal complications (Badillo et al. 2007; Davis et al. 2009). Factors, such as intrauterine growth restriction, thickened bowel, and stomach herniation, have also been proposed as parameters predicting poor postnatal outcomes. There are two commonly accepted explanations for the thickened, foreshortened, and edematous bowel (D'Antonio et al. 2015; Horton et al. 2010). Continuous contact of the herniated contents with amniotic fluid has been proposed as one reason. Secondly, serial fetal ultrasonography has demonstrated that the fascial defect begins to decrease in diameter near the end of the third trimester, which could lead to venous congestion, swelling, and even infarction of the midgut. Therefore, serial fetal ultrasonography in the third trimester is warranted to follow the appearance of the bowel. Progressive worsening of these findings could lead to future complications

and warrant early delivery. However, the timing of delivery still remains controversial. Rationale for early delivery (36–38 weeks) includes decreased exposure time of the intestine to amniotic fluid and ability to ensure delivery at/near a tertiary care pediatric center. However, advocates for spontaneous delivery believe that early delivery is associated with poorer outcomes such as increased ventilation requirements from lung prematurity, prolonged time to full enteral feeds, and prolonged hospital stay. The findings from a recent study indicate that there is a decreased incidence of severely matted bowel with increased gestational age, further discouraging early or preterm delivery (Youssef et al. 2015). It has been demonstrated by numerous studies that the mode of delivery (vaginal vs. Cesarean section) does not influence patient outcomes in gastroschisis (Snyder et al. 2011). Delivery at or near a tertiary pediatric facility, with the availability of a pediatric surgeon and level III neonatal intensive care unit, has been shown to significantly improve outcomes for these patients (Savoie et al. 2014). These infants are likely to have more prompt surgical intervention and successful primary closure. This is likely due to shorter exposure time of the herniated bowel resulting in decreased swelling and perhaps the need for less bowel manipulation. These factors likely lead to earlier initiation of enteral feedings and decreased lengths of stay.

Preoperative Assessment and Preparation

Infants with gastroschisis require prompt intervention. Significant delays in management of the herniated contents should be avoided. Appropriate preoperative preparation is essential to ensure a good outcome. Because heat loss from the exposed herniated contents can be significant, maintenance of the infant's temperature within the normal range is critical. It is well documented that hypothermia leads to poorer overall outcomes with delayed bowel function and prolonged length of stay. There are various

methods to minimize this heat loss depending on available supplies in the delivery room or intensive care nursery. If transport of the infant to a pediatric surgical center is needed, the patient should have an intravenous catheter established, the bowel should be wrapped in moist saline-soaked gauze, and the lower body placed in a plastic bag to the lower chest or loosely wrapped in cellophane. Alternatively, dry rolls of gauze may be wrapped around the patient's abdomen after the damp gauze-covered bowel is placed to create an appropriate environment. It is imperative to stabilize the bowel to diminish the risk of compromising its blood supply at the fascial ring. The infant should be in a warming isolette or under an overhead radiant warmer to help maintain normothermia.

Most infants with gastroschisis are dehydrated at birth and require at least 125% of normal maintenance fluids to regain normovolemia. Eventually, almost all infants with gastroschisis will require central venous access. Broad-spectrum antibiotics are appropriate during the perioperative period because of exposure of the bowel and peritoneal cavity to bacterial contamination at the time of birth. A nasogastric or orogastric tube placed at the time of birth is necessary for gastrointestinal decompression because of the bowel inflammation and resulting ileus. Thus, before surgery, the infant with gastroschisis should be normothermic, normovolemic, and hemodynamically stable and have normal serum electrolytes following adequate fluid resuscitation.

Anesthesia

General anesthesia is required for appropriate operative management of gastroschisis. The choice of anesthetic agents should be made by the anesthesiologist, but there are two important considerations: (1) muscle paralysis is useful in optimizing the chances for complete reduction of the herniated bowel and primary abdominal wall closure; and (2) nitrous oxide should not be used

as it diffuses into the lumen of the bowel causing distension and compromises the likely success of primary abdominal wall repair.

Operative Procedure

The operation should be performed under conditions that maintain normothermia. Several methods exist to accomplish this goal. An overhead radiant warmer, warming lights, or a warming blanket should be used to maintain the infant's temperature in the normal range during the procedure. Raising the temperature in the operating room may also be necessary. After the induction of general anesthesia, the dressing previously placed over the herniated contents should be removed. The bowel should be handled with sterile gloves. The umbilical cord, which has usually been left long, should be clamped 2–3 cm above the abdominal wall and the excess cord then removed. Holding the bowel and clamp on the umbilical cord in one hand, the bowel should be prepared using gauze sponges soaked in a 50:50 mixture of povidone-iodine solution and saline. The antiseptic solution must be warm to the touch in an effort to minimize heat loss. After gently washing the bowel and the anterior and lateral abdominal wall, drapes are appropriately placed and the herniated contents are laid on the drapes. The surgeon should then scrub and put on gown and gloves.

At this point the umbilical stump can be ligated allowing removal of the clamp. Next, the herniated intestine should be carefully inspected for areas of perforation or sites of atresia, although no effort should be made to dissect matted loops of intestine. It is sometimes necessary to extend the abdominal wall defect to facilitate reduction of the herniated bowel. This is generally done by extending the defect superiorly in the midline by 1–3 cm (Fig. 3). Extending the incision caudally is not recommended because the urinary bladder is in close proximity to the inferior aspect of the abdominal wall defect. The length of this incision depends on the size of the original defect and the bulkiness of the herniated bowel.



Fig. 3 Vertical extension and closure of gastroschisis defect

Primary Closure

The herniated intestine is reduced as much as possible, distributing the bowel to all quadrants of the peritoneal cavity. Two techniques have been described to facilitate complete bowel reduction and abdominal wall closure: (1) stretching of the anterior abdominal wall and (2) “milking” the intestinal contents into the stomach and aspiration through the nasogastric tube. Some surgeons also find milking out the colonic contents to be an effective maneuver for decompressing and reducing the bowel. Although gentle stretching of the anterior abdominal wall can be useful, vigorous stretching can lead to rectus muscle hemorrhage and abdominal wall edema, producing a non-compliant, firm anterior abdominal wall. This can result in ventilation difficulties and wound-related problems. Caution should also be taken when manipulating the intestine to “milk” the intestinal contents into the stomach, as this can cause further damage to the bowel wall, resulting in increased bowel wall thickening and additional delay in bowel recovery. If reduction of the herniated intestine is successful, the abdominal wall is assessed for primary closure. If it can be closed without undue tension, 3/0 absorbable, monofilament sutures are used. These sutures are placed in a figure-of-eight fashion, as this results

in fewer knots and greater distribution of tension. When all the sutures have been placed, they are tied in sequence with a thin, malleable retractor initially underneath the fascia to prevent a loop of intestine from becoming entrapped under the sutures. The umbilical stalk is retained to create a more natural umbilical appearance when the wound is fully closed. When the fascia has been closed, the skin edges are approximated using interrupted absorbable sutures or skin staples and sufficient sterile skin closure strips, allowing distribution of skin tension over a wider surface area and thus reducing the likelihood of skin disruption.

About 60–70% of infants with gastroschisis can be operatively treated in this way without creating undue intra-abdominal pressure or tension in the abdominal wall closure. It is best to avoid high intra-abdominal pressure and excessive suture line tension. This can result in abdominal compartment syndrome, possibly leading to intestinal necrosis, renal hypoperfusion, and difficulty in ventilation, as well as wound disruption. Intra-gastric and bladder pressure monitoring has been used by some pediatric surgeons to determine intra-abdominal pressure (Lacey et al. 1993). These two measurements are used as a guide to monitor intra-abdominal pressure during primary or staged closure of gastroschisis. The goal of therapy is to maintain intra-abdominal pressure below 20 mmHg, which is based on prior studies showing that higher pressures compromise intra-abdominal organ perfusion.

An alternate method, utilizing abdominal wall component separation, has been described for closure of larger gastroschisis fascial defects (Levy et al. 2013). This technique has been used for many years to repair large ventral hernias in older children and adults. It involves incising the external oblique fascia lateral to the rectus sheath and separating the external oblique from the internal oblique muscle. This is done bilaterally and allows for tissue approximation in the midline without significant tension. Biologic mesh may be placed above or below the tissue to reinforce the repair if needed.

Staged Closure

For patients in whom complete reduction of the herniated bowel and abdominal wall closure are not possible or appropriate, the staged reduction technique described by Schuster in 1967 has proved to be very useful (Schuster 1967). Reinforced Silastic sheeting (0.8–1.0 mm thick) is sutured to the fascial edges (Fig. 4). This is accomplished with interrupted 3/0 silk mattress sutures. It is generally necessary to enlarge the fascial defect prior to suturing the Silastic sheet. However, extending the fascial opening too far inferiorly should be avoided as bladder injury may occur. The cephalad and caudad vertical edges of the silo are constructed with running 3/0 monofilament sutures (Fig. 5). Before closing the top of the silo, as much of the bowel as possible is reduced into the peritoneal cavity by manual compression within the sac while avoiding excessive intra-abdominal pressure. The top of the sac is oversewn with a 3/0 monofilament suture placed in a running horizontal mattress

s fashion. Suture is also placed through the skin and looped over the silo (Fig. 6) in order to place some tension on the skin edges to minimize skin retraction and facilitate skin approximation at the time of the abdominal wall closure. The silo is covered with povidone-iodine ointment followed by dry roll gauze to act as a protective dressing and provide support to the silo at the fascial level. The staged reduction technique requires daily reduction of the herniated intestine within the

silo. The optimal target for completely reducing the bowel, removing the silo and closing the abdominal wall, is 7–10 days. Any delay beyond this timeframe substantially increases the risk of fascial infection, tearing away of the silo from the anterior abdominal wall muscle, and failure of the technique. The risk of failure of this technique is high if the silo is not able to be removed within 14 days from placement. Daily reduction of the intestinal contents within the sac can be accomplished in the neonatal intensive care unit using sedation and sterile technique (Fig. 7). Each time



Fig. 5 Silo creation using Silastic sheeting

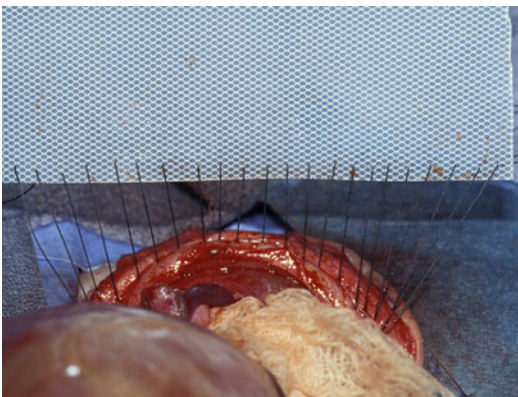


Fig. 4 Silastic sheeting sutured to fascial edge

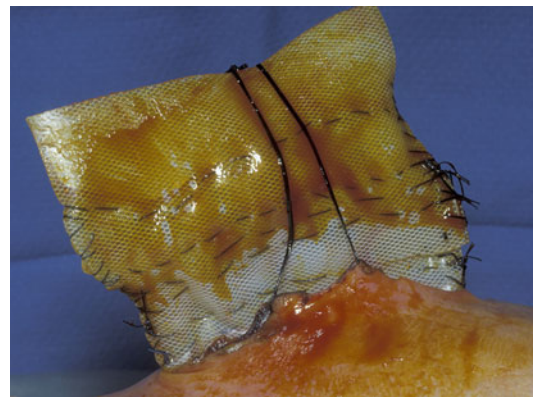


Fig. 6 Suture placed through skin and looped over silo

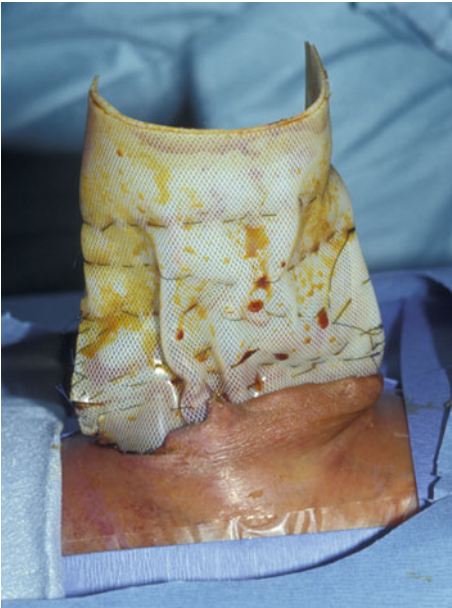


Fig. 7 Reduction of bowel in silo using horizontal suture

the procedure is performed, the sac and anterior abdominal wall are prepared with warm povidone-iodine solution before the reduction, and povidone-iodine ointment is applied followed by roll gauze after the procedure. General anesthesia is not necessary. When the herniated bowel has been successfully reduced into the peritoneal cavity and the fascial edges brought to within 1 cm of each other, the infant is ready for removal of the sac and primary abdominal wall closure in the operating room under general anesthesia (Schwartz et al. 1983).

An alternative method of staged reduction which has become popular and is effective is the placement of a preformed, spring-loaded silo at the bedside (Fig. 8) (Schlatter et al. 2003). This can be accomplished without general anesthesia. The preformed silo comes in different diameters and should be selected appropriately to accommodate the size of the defect but more importantly to accommodate the bulkiness of the herniated contents. The ring of the spring-loaded silo is placed underneath the fascial defect after the herniated bowel is placed within it (Fig. 9). A very small fascial defect can be constrictive and may lead to failure to reduce the herniated contents into the peritoneal cavity. Also, placing the bowel

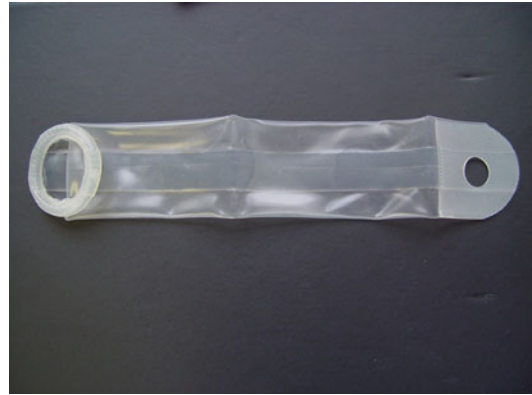


Fig. 8 Preformed, spring-loaded silo



Fig. 9 Bowel placed within preformed silo

contents into a small diameter preformed silo can result in local or massive intestinal ischemia and/or infarction. If the defect is too small, the fascial opening should be enlarged to allow use of a larger diameter silo to prevent these potential complications. Reduction of the bowel is accomplished in a similar fashion to that used for the sutured silo, except that a single tie with umbilical tape is used to secure the reductions (Fig. 10).

Recently, another method, referred to as “sutureless” or “plastic” gastroschisis closure, has been described (Sandler et al. 2004). In this technique, the bowel is reduced in the usual fashion either primarily or after placement in a silo. However, instead of placing sutures to approximate the fascia, the defect is covered with the umbilical stump or a nonadherent dressing. An occlusive



Fig. 10 Reduction of abdominal contents within spring-loaded silo

dressings is then placed over the site and the wound is allowed to granulate. Once granulation tissue covers the wound bed, the area is covered with dry dressings. Proposed advantages of this technique include reduced intra-abdominal pressure during the closure process and decreased narcotic and sedation requirements. Nearly all infants have an umbilical hernia following this method of repair, but many of these resolve spontaneously, similar to isolated umbilical hernias (Riboh et al. 2009; Orion et al. 2011; Choi et al. 2012).

Negative pressure wound therapy has also been utilized to manage very large abdominal wall defects. This technique can be used for initial coverage of viscera in patients without sufficient abdominal domain. This negative pressure dressing can also be placed above a skin graft or over a closed wound to facilitate granulation tissue and decrease tension on the repair site. The exact mechanical settings are variable based on patient status and goals of therapy (McBride et al. 2014).

Postoperative Care

Patients with gastroschisis require parenteral nutrition to provide the necessary calories intravenously while awaiting bowel reduction and recovery of bowel function. This can be accomplished via a cuffed Silastic central venous catheter or a peripherally inserted central catheter (PICC line). Parenteral nutrition is typically required for 2–4 weeks

while awaiting the return of intestinal function. Nasogastric decompression is necessary until there is evidence of bowel function. Broad-spectrum antibiotics are generally continued during the perioperative period (usually 3–5 days). Those infants who undergo the staged approach require a longer period of antibiotic treatment (usually until 1–2 days after the silo has been removed). Once there is evidence of gastrointestinal function, enteral feeding can be introduced and gradually progressed using breast milk or a low-residue elemental-type formula with appropriate caloric intake. In the past, enteral feeding in these patients was generally delayed for at least 4–6 weeks after surgery, as it was thought that early feeding could lead to an increased risk of developing complications. However, this approach has not been supported by clinical evidence. More recently, enteral feedings have been started as early as 10–14 days after abdominal wall closure with no increase in adverse outcomes.

Complications

Complications in infants with gastroschisis are generally related to the gastrointestinal tract or the abdominal wall closure. As noted earlier, in utero complications from intestinal atresia or perforation can occur (Fig. 11). Intestinal perforation can be managed in one of several ways, depending on the specific circumstances. The options at the time of birth include suture closure (Fig. 12), resection of the site of perforation with oversewing of the two ends of the bowel (i.e., creating “intestinal atresia”), or creation of a stoma if primary abdominal wall closure can be accomplished. It is generally not recommended to attempt a bowel anastomosis because of the marked thickening and inflammation of the bowel wall.

Intestinal atresia (Fig. 13) can be managed by the creation of a stoma if primary abdominal wall closure is possible or by leaving the atresia in situ if staged reduction is undertaken. A stoma can be created at the time of removal of the silo and primary abdominal wall closure. Alternately, the atresia can be left in place at the time of the abdominal wall closure, especially in cases where significant intestinal wall thickening and



Fig. 11 Multiple intestinal perforations in gastroschisis bowel

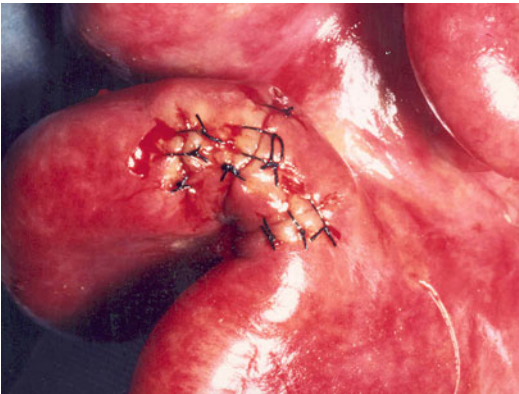


Fig. 12 Suture closure of intestinal perforations



Fig. 13 Intestinal atresia in gastroschisis

inflammatory peel is present. The patient will then undergo a re-exploration in 4–6 weeks with definitive surgical management of the atresia at that time (Snyder et al. 2001).

A devastating complication of gastroschisis can be partial or complete necrosis of the midgut as a result of excessive intra-abdominal pressure or kinking of the blood supply to the bowel before or at the time of reduction of the herniated bowel. This complication may lead to the death of the patient or to short bowel syndrome. Thus, excessive tension creating increased intra-abdominal pressure should be avoided to minimize this complication. Additional complications associated with the abdominal wall closure are wound dehiscence and intestinal-cutaneous fistula formation. These complications are also often associated with excessive intra-abdominal pressure. Therefore, it is preferable to use the staged reduction approach when primary abdominal wall closure might result in excessive intra-abdominal pressure (Kunz et al. 2013).

A delayed complication is the development of necrotizing enterocolitis. The incidence of necrotizing enterocolitis in patients with gastroschisis has been reported to be as high as 20% (Oldham et al. 1988). It generally has a delayed onset, usually 3–6 weeks after birth. The causes remain unknown, but associations have been made with total parenteral nutrition (TPN)-induced cholestasis and slow recovery of bowel function. Necrotizing enterocolitis associated with gastroschisis can be mild or severe and can involve a significant portion of the bowel resulting in a high mortality. Finally, sepsis, resulting from intra-abdominal or wound infections and central line infections, are additional causes of morbidity in the gastroschisis patient (Youssef et al. 2017).

Feeding intolerance is a common finding in gastroschisis patients and is typically a result of gastroesophageal reflux and intestinal dysmotility. Both of these can usually be managed medically, with adjustment of feeds and/or medications, including H₂-blockers, proton pump inhibitors, and prokinetic agents. In cases of severe gastroesophageal reflux where medical management is insufficient, anti-reflux surgery may be necessary.

A recent study from one institution suggested that there may be a higher incidence of hiatal hernias in gastroschisis patients and this, in turn, may contribute to more severe reflux requiring surgical intervention (Tsai et al. 2014).

Outcome

The availability of neonatal intensive care units, parenteral nutrition, and the technique of staged reduction have resulted in significant improvement in the outcome for infants with gastroschisis over the past four decades. The survival of infants with gastroschisis has exceeded 90% (Lap et al. 2016). Morbidity should be relatively low if attention is paid to the details of the surgical correction. In addition to the marked improvement in survival, the lengths of time to initiation of feedings and hospital discharge have been significantly shortened. Whereas hospitalization times usually exceeded 6 weeks, the average is now around 4 weeks (Kong et al. 2016, Puligandla et al. 2017). Infants successfully treated for gastroschisis typically do not have significant complications during later infancy and childhood. Several studies have shown that their growth and development is normal or near normal. The need for later surgical intervention is usually for bowel obstruction (~8%). Other surgical procedures included abdominal wall hernias and scar revisions.

Conclusion and Future Directions

The successful management of infants with gastroschisis has evolved over the last four decades with advances that have been made in perioperative management and surgical techniques. Future research could focus on prenatal evaluations and evolving surgical techniques. Further studies are needed to determine which prenatal parameters correlate with poor postnatal outcomes. Also, more long-term data is needed for evaluation of sutureless closure techniques and outcomes.

Cross-References

- ▶ [Embryology of Congenital Malformations](#)
- ▶ [Fetal Counseling for Congenital Malformations](#)
- ▶ [Long-Term Outcomes in Newborn Surgery](#)
- ▶ [Omphalocele](#)
- ▶ [Prenatal Diagnosis of Congenital Malformations](#)

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