# Gastroschisis

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Saleem Islam

Gastroschisis and omphalocele are the two most common congenital abdominal wall defects. Gastroschisis is the more common of the two with an increasing incidence worldwide. Embryologically, the intestine herniates into the umbilical cord region from the sixth to the tenth week of gestation due to rapid elongation and growth of the abdominal viscera. The etiology of gastroschisis is not entirely clear; however the most accepted theory suggests that there is failure of migration of the lateral embryonic folds, which is more common on the right side, resulting in a defect to the right of the umbilical cord. This implies that the defect occurs early during gestation.

The incidence is increasing worldwide and is approximately 1:4000 live births. In the USA the incidence increased from 2.3 per 10,000 live births in 1995 to 4.4 per 10,000 in 2005. Tobacco use during pregnancy, a variety of common medications (aspirin), and insecticide contamination of the soil have all been implicated but without conclusive evidence. There is a strong association with young maternal age. Almost 40 % of gastroschisis cases occur in mothers less than 21 years age. Low socioeconomic status, illicit drug use, and low body mass index have also been suggested as contributory. Race may also play a role, as the incidence is lower in African Americans while a higher incidence is seen in Hispanics.

## Prenatal and Perinatal Management

Prenatal diagnosis for gastroschisis occurs in the majority of cases. An elevated maternal serum alpha-fetoprotein (AFP) may be the earliest sign. Up to 90 % of the defects are picked up on prenatal ultrasonography by the mid second trimester. Detection of free-floating loops of intestine in the amniotic fluid to the right of the umbilical cord is pathognomonic. Prenatal detection of an abdominal wall defect typically leads to a transfer in care of the pregnant patient to a high-risk center. Amniocentesis is not considered to be of benefit in isolated gastroschisis, but is used if additional defects are present.

Intestinal damage is seen in many cases of gastroschisis. This ranges from an inflammatory peel or serositis to intestinal necrosis or perforation. The inflammatory injury to the bowel does not occur in all cases and the severity is variable. The etiology is unknown but is thought to be the result of exposure to the amniotic fluid and constriction of the bowel with lymphatic obstruction at the abdominal wall. Waste products including urea, cytokines, meconium, and other substances have been implicated in the inflammatory response resulting in deposition of type IV collagen, bowel wall thickening, and intestinal dysmotility. Animal research in which amniotic fluid is exchanged for lactated ringers has resulted in improvement in the inflammatory response and decreased thickening of the bowel. Human trials with amniotic fluid exchange have been inconclusive.

Bowel diameter on prenatal ultrasonography has been evaluated as a potential marker for complex gastroschisis. The average bowel diameter prior to 30 weeks gestation is <2 mm. Bowel dilation >10 mm prior to 30 weeks gestational age has been predictive of atresia while intestinal damage may be seen in 35–40 % of neonates with bowel dilation ≥6 mm. Attempts have been made to correlate fetal US findings with postnatal outcomes. Intra-abdominal bowel dilation is significantly associated with bowel compromise, while bowel wall thickening, extra-abdominal dilation, or gastric dilation is not. Overall, fetal US measurements of bowel diameter and thickness are inconsistent predictors of clinical outcome and need further refinement.

Preterm delivery to limit intestinal exposure to the amniotic fluid has been proposed for gastroschisis. In theory, limited exposure would result in less injury thereby increasing the chance of early abdominal wall closure, improving intestinal motility, and decreasing the time to initiate feeds.

S. Islam, MD, MPH (✉)
Department of Pediatric Surgery, University of Florida College of Medicine, 1600 SW Archer Road, PO Box 100119, Gainesville, FL 32610, USA
e-mail: saleem.islam@surgery.ufl.edu



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Fetuses with gastroschisis are delivered prematurely more often (25–30 %) than those without abdominal wall defects (~6 %). Management protocols that include planned cesarean section between 36 and 38 weeks and as early as 34 weeks have resulted in high rates of primary repair. However, delivery before 37 weeks is associated with increased time to full enteral feedings and increased length of stay compared to term delivery. Furthermore, infants with gastroschisis who weigh <2 kg experience prolonged hospitalization, more ventilator days and increased time to full feeds. Given the added risks to the neonate and mother with planned preterm cesarean section, the evidence favors term delivery.

The mode of delivery for the fetus with gastroschisis is often a point of discussion. Some advocate routine cesarean section in order to avoid trauma to the exposed bowel from vaginal delivery and avoid exposure of the bowel to the vaginal flora. However, no clear benefit to cesarean delivery has been demonstrated. Therefore, a vaginal delivery at term is generally recommended unless contraindicated secondary to maternal or fetal issues. Many advocate delivery of the fetus with gastroschisis at a specialty center with full-time pediatric surgical and neonatal capabilities.

## Neonatal Resuscitation

Newborns with gastroschisis experience considerable evaporative fluid loss. Immediately following delivery the patient's airway, breathing and circulation should be evaluated then attention turned toward management of the gastroschisis. Endotracheal intubation is used only if indicated for respiratory distress. Orogastric or nasogastric decompression is performed. Vascular access is obtained and intravenous fluids and antibiotics started. An initial isotonic fluid bolus of 20 mL/kg is given. In the past, 1.5–2× "maintenance" fluids have been recommended for the initial resuscitation. Cautious preclosure fluid resuscitation is now encouraged as excessive fluid may result in adverse outcomes such as increased ventilator days, days of parenteral nutrition, length of stay, and occurrence of bacteremia. The bowel is quickly inspected for signs of ischemia or a tight fascial ring then covered with a plastic bag over the torso ("bowel bag") to reduce fluid losses for transport to the NICU. After the bowel is covered, the baby is placed right side down and the gastroschisis stabilized to avoid kinking of the vascular pedicle. The use of narcotic pain medication at this stage is not required, however, some neonatal units prefer to use a short acting drug such as Fentanyl. Emergent surgical evaluation is necessary. If this is not available, transfer to a facility with surgical expertise must be arranged.

## Surgical Care

Surgical evaluation of the bowel is rapidly performed to ensure there is no kinking of the blood supply related to patient position or constriction of the blood supply related to a small fascial defect. If a constricting defect is identified, the defect is opened at the fascial level, usually on the right lateral position. In addition to the intestine and stomach, care must be taken to look for intestinal atresia or other herniated structures. The gonads (ovary or testicle) may be herniated through the defect, and should be carefully placed into the abdominal cavity. Occasionally, the top of the bladder is noted to be protruding from the bottom of the defect. While an atresia may be suspected on initial inspection, it is often unclear in cases with extensive peel (Fig. 68.1). Similarly, the intestine may appear to have inadequate length initially due to the inflammation. Any bands crossing the bowel loops should be divided prior to placing a silo or primary abdominal wall closure.

Controversy exists over the closure technique for gastroschisis. Before the early 1990s, the standard was to attempt primary closure in all cases, and then construct a silo if closure was not possible. The introduction of the preformed silastic silo with a spring-loaded ring changed the paradigm, as it could be placed at the bedside with minimal sedation and avoids intubation. This technique was rapidly adopted across the USA and throughout the world.

## Primary Closure

This technique involves immediate reduction of the herniated viscera to the abdominal cavity and closure of the defect.

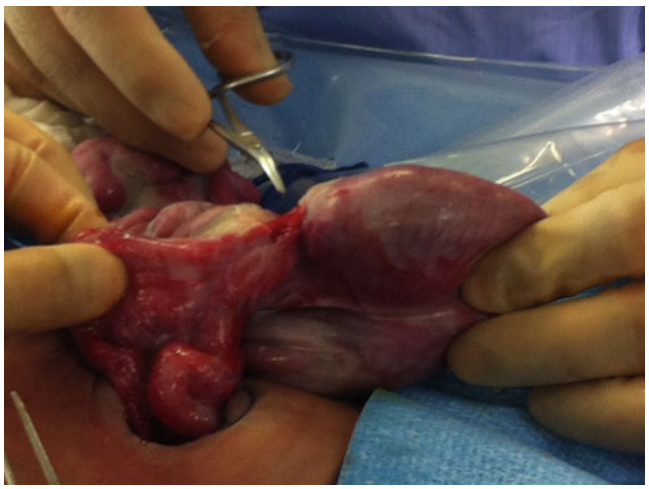

**Fig. 68.1** Gastroschsis initial evaluation. Note the extensive inflammatory peel and foreshortened mesentery

Success rates for primary closure as high as 80 % have been reported. The procedure has been performed at the bedside, but more often is done in the OR under general anesthesia. The first step involves assessing the degree of inflammation and edema and searching for an atresia. A vigorous effort is then made to empty the large intestine and rectum of meconium performing irrigations with a catheter placed through the anus if necessary. Care must be taken to ensure there is no serosal injury. The abdomen is then stretched manually with the surgeon's fingers to create additional room, and the intestine then returned into the abdomen.

Various methods have been described to measure the abdominal pressure during this part of the procedure; including intra-gastric pressure, urinary bladder pressure, and peak inspiratory pressure on the ventilator. Intra-abdominal pressures >20 mmHg are associated with decreased visceral perfusion and an increase in peak inspiratory pressure >10 mmHg suggests a significant increase in intra-abdominal pressure. Once the contents are returned to the peritoneal cavity, the defect itself can be closed in a variety of ways. Fascial closure may be performed followed by skin closure and umbilical reconstruction by preserving the base of the cord. This technique results in a higher intra-abdominal pressure; so if there is concern about the pressure, skin closure alone can be performed. Subsequent spontaneous closure of the fascial defect similar to an umbilical hernia is anticipated in many of cases following skin closure alone. Finally, bedside reduction followed by "sutureless" repair using part of the umbilical cord to plug the defect has been successful.

## Silo and Delayed Closure

Schuster first described the use of a silastic sheet sewn to the skin and fascia to create a silo in a neonate with gastroschisis. The preformed silo was introduced in the 1990s and became rapidly accepted, consisting of a spring-loaded silastic covered ring that was inserted into the abdominal cavity beneath the fascia with a transparent external silo (Fig. 68.2). The device comes in a number of sizes based on the diameter of the ring (3–10 cm). The ring is pinched and inserted into the abdominal cavity after the intestines have been gently placed into the silo (Fig. 68.3). The bowel is then serially reduced over the next 2–5 days using clamps or twisting and ligating the silo with an umbilical tie. Some authors advocate allowing the bowel to spontaneously reduce into the abdominal cavity and once a plateau is reached then perform definitive closure.

A major advantage of the silo technique is the gentle reduction of the viscera over time in order to decrease the effects of increased abdominal pressure. Reduced need for mechanical ventilation, reoperation for intra-abdominal hypertension, and incidence of necrotizing enterocolitis have been reported with silo use. In addition, silo placement allows for an elective procedure to achieve definitive closure. Disadvantages include increased incidence of sepsis, delay in closure, and increased time to initiation of feeds.

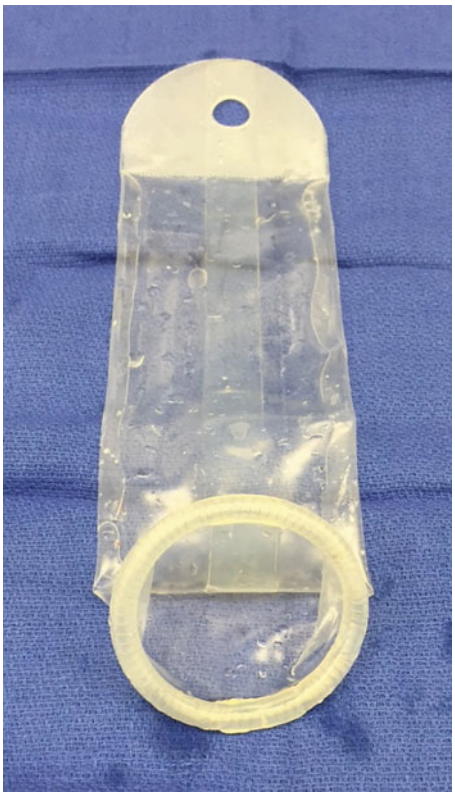

**Fig. 68.2** Preformed spring-loaded silastic silo

## Complicated Gastroschisis

Some newborns with gastroschisis will have associated intestinal atresia, intestinal perforation, prenatal loss of intestine with resultant short bowel (vanishing midgut), or bowel ischemia. These patients have overall worse outcomes and risk adjustment models have noted that long-term morbidity and complications are almost entirely in this group. Complex defects account for 5–15 % of cases in most reported series and result in a longer hospital length of stay, increased use of parenteral nutrition and parenteral nutrition associated cholestasis, higher readmission rates, increased total cost, and higher mortality rates compared to the simple gastroschisis cohort.

Intestinal atresia is noted in approximately 10–15 % of gastroschisis patients and management of these cases is individualized according to the location of the atresia and severity of the peel and inflammation (Fig. 68.4). If there is minimal bowel matting and thickening, an atresia may be safely primarily repaired at the time of abdominal wall

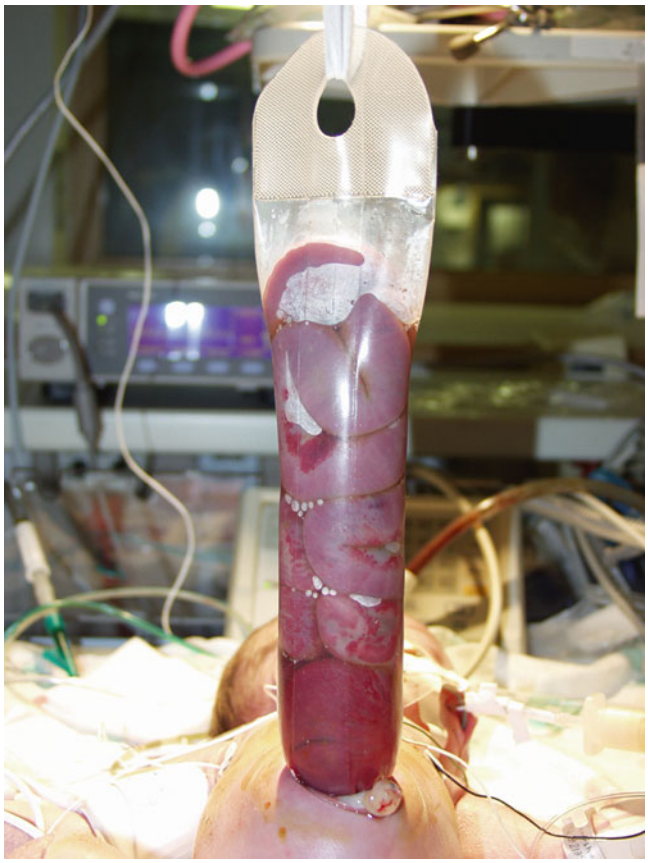

**Fig. 68.3** Appropriate placement of silo. Bowel is placed in the smallest size silo that will accommodate it. Viable and well-perfused bowel visible within silo without much bottle-necking at level of defect

closure. However, if there is significant inflammation of the bowel wall, reduction of the bowel and defect closure can be performed and repair of the atresia delayed 4–6 weeks when the inflammatory peel has resolved. In cases of a distal intestinal atresia, the creation of a stoma is preferred to allow early enteral feeding and prevent continued distention of the proximal segment.

## Special Considerations

Closed or vanishing gastroschisis is a rare condition in which the defect either completely or partially closes in utero, resulting in strangulation of the herniated bowel. This may present as a newborn with necrotic herniated midgut or as a newborn with a closed abdominal wall and limited intestinal length. These patients have short bowel syndrome requiring long-term parenteral nutrition and possibly bowel lengthening procedures or small intestine transplantation.

Ruptured omphalocele is another rare scenario that bears mentioning as it may be confused as a gastroschisis. In these cases, liver is included in the herniated viscera and the defect is much larger than the usual gastroschisis opening (Fig. 68.5). Management of these neonates is complex and requires staged closure, with outcomes much worse than a standard gastroschisis.

## Outcomes

Gastroschisis outcomes have dramatically improved over the past four decades. Mortality was close to 50 % in the 1960s and currently is less than 10 %. Survival in simple gastroschisis is over 95 %, while 80–90 % survival is noted in complex cases.

Morbidity has also similarly improved with time. Most of the morbidity in gastroschisis is associated with dysmotility of the gastrointestinal tract. This is significantly worse in complex cases. Prolonged use of parenteral nutrition in patients who are unable to feed may result in cholestasis, and in some cases liver failure. The use of motility agents has not been beneficial in improving bowel function. However, clinical management pathways have resulted in decreased time to full enteral feeds. In patients who do not have bowel function or who are unable to tolerate enteral feeds within 3–4 weeks after closure, an intestinal atresia should be sought. Pneumatosis intestinalis and necrotizing enterocolitis occur in up to 10 % of neonates with gastroschisis but most will respond to non-operative management. Length of stay is close to 4 weeks on average and is generally longer in complex cases.

All patients with gastroschisis have malrotation. A Ladd procedure is usually not performed, as adhesions from the repair are presumed to likely prevent a future volvulus. The family should be alerted to the significance of bilious emesis and encouraged to seek prompt intervention. Other anomalies such as undescended testes are noted in 35–40 % of boys. In cases where the testicle is herniated through the defect, initial management consists of simple reduction of the gonad. Approximately half of patients with cryptorchidism will experience testicular descent. At a year of age, those with an undescended testicle should undergo an orchiopexy. If the testicle is non-palpable, it is reasonable to attempt diagnostic laparoscopy to evaluate the testicular position.

Children with gastroschisis are typically small for gestational age but will usually catch up to their peers by 12–24 months of age. However, outcomes data suggests that long-term neurodevelopmental outcomes are lower than age-matched cohorts. Additionally, roughly 60 % of patients report being dissatisfied with the appearance of their umbilicus, which can also result in psychosocial stress.

## Care Management Pathway

The mother of a fetus with gastroschisis is referred for prenatal consultation. The anticipated prenatal, perinatal and postnatal course as well as potential complications and surgical management are discussed. The prospective parents also tour

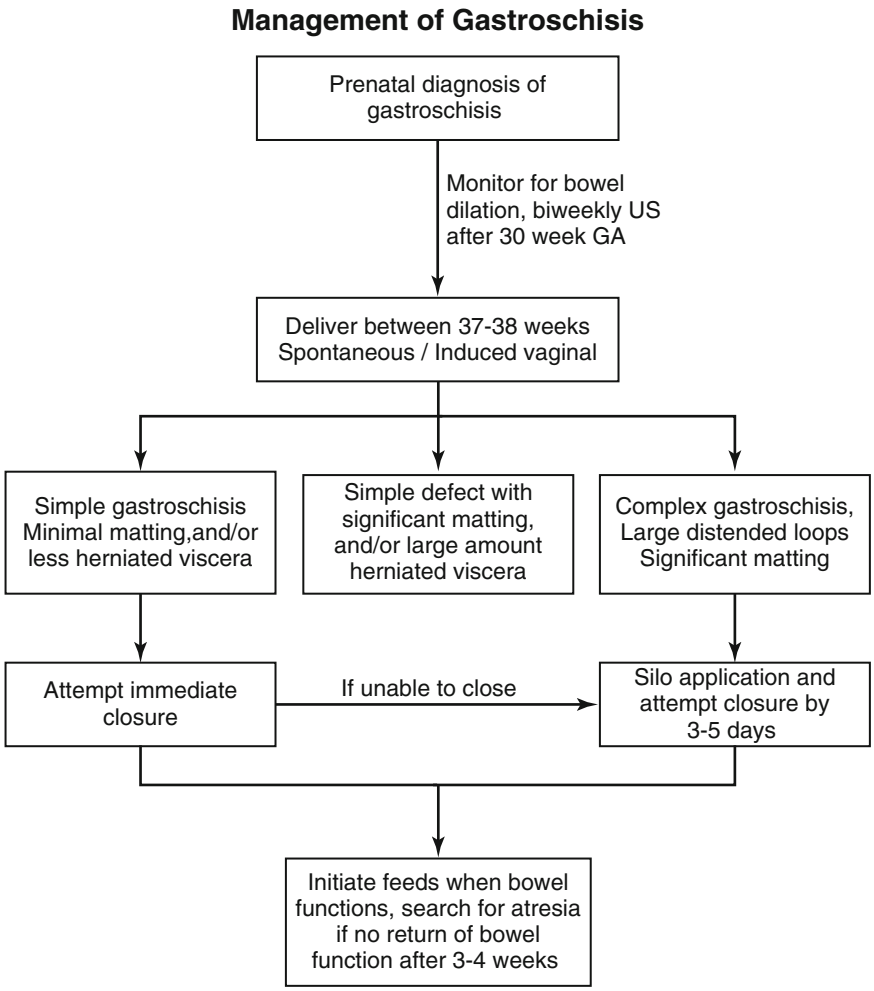


**Fig. 68.4** Suggested algorithm for gastroschisis management

the NICU and discuss issues with the nurses and social workers. This helps to alleviate many concerns and prepare them for the postnatal care.

Communication with the high-risk maternal fetal medicine specialists allows for a coordinated delivery with pediatric surgical presence in the NICU to immediately receive the neonate. As resuscitation is proceeding, the gastroschisis defect and intestine are examined. I prefer to place a spring-loaded silo at the bedside for most cases and reserve immediate closure for cases with relatively little herniated and inflamed viscera. Twice-a-day reductions are started 12–24 h after placement of the silo and a PICC line placed for parenteral nutrition. The goal in all cases is to have the viscera reduced by 3–5 days and perform a final closure in the operating room. I prefer to close the skin only and have had good cosmetic results. Most of the fascial defects will spontaneously close by 3 years of age. Antibiotics are administered while the silo is in place and continued for 24 h after closure. If an atresia is identified, I perform an ostomy at the umbilicus.

An orogastric tube is kept on suction until the volume diminishes to less than 15–20 cc per day and then the tube is taken off suction and removed if tolerated. Feeding is initiated via continuous feeds through a small bore feeding tube to allow for gut priming and then advanced if tolerated and bowel function begins. For simple gastroschisis a hospital stay of 3–4 weeks is expected. Patients are followed for at least 2 years after discharge to ensure adequate weight gain and that there are no feeding issues.

## Editor's Comment

The delivery of a fetus with gastroschisis should be performed at a center all the resources needed for definitive care. Labor may be spontaneous or induced as close to term as possible. Cesarean section delivery should be reserved for the usual maternal and fetal indications.

Following delivery, the intestines should be covered with a bowel bag and the patient placed in a right lateral position to avoid kinking of the mesentery during transport. Although no closure technique is clearly superior, bedside placement of a spring-loaded silo allows for rapid coverage of almost all defects in order to prevent insensible water losses and protect the bowel. Serial reduction of

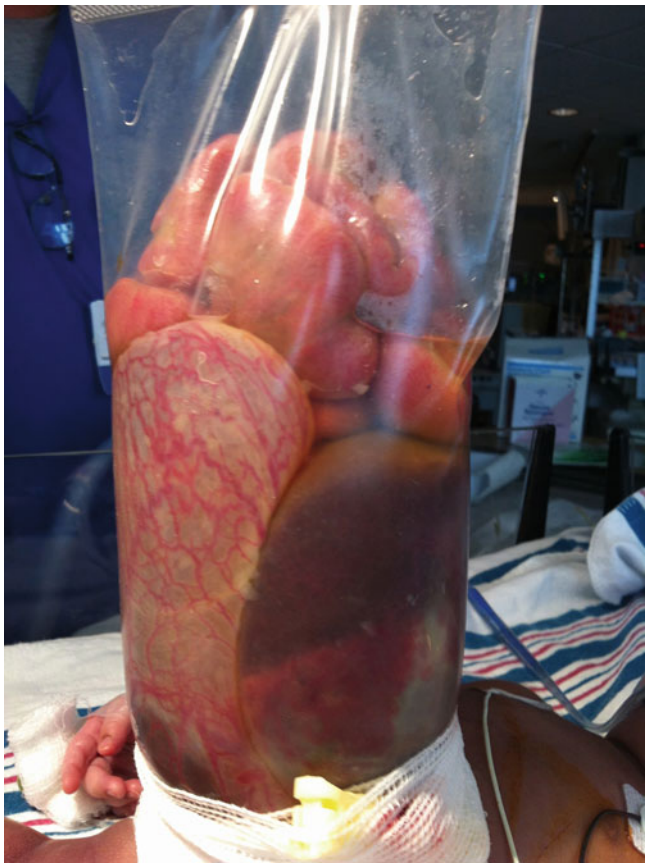

**Fig. 68.5** Ruptured omphalocele. Note the large amount of herniated liver in the silo. Closure was eventually performed with a skin graft covering the bowel and the ventral hernia later repaired

the gastroschisis should be performed one or two times a day with a goal of complete reduction within 3–5 days. If the silo is left in place for longer periods of time, the fascial edges have a tendency to recede making the definitive fascial closure more difficult or a prosthetic patch necessary to avoid intra-abdominal hypertension with closure. The best intra-operative sign of excessive pressure is the peak inspiratory pressure. A number of techniques for fascial and skin closure have been described. A vertical midline closure of the fascia after creation of generous skin and subcutaneous flaps and creation of a neoumbilicus by marsupializing the skin edges to the fascia at the desired location and appropriate size for an umbilicus provides good cosmesis.

Antibiotics are usually continued postoperatively for 48 h and the patient maintained on parenteral nutrition until non-bilious, low-volume gastric output, and bowel function. Feedings are then slowly advanced. It is not uncommon for patients to become intolerant of feeds after several days of advancement. The feeds can be held briefly or decreased and then advancement resumed as long as there are no signs of sepsis and an abdominal radiograph shows no pneumotosis intestinalis. If the patient did not pass meconium preoperatively and has not had bowel function within 4 weeks following closure, an atresia or stricture should be ruled out. A contrast enema is obtained initially and if normal an upper GI performed after the contrast from the enema has cleared.

Management of an atresia diagnosed on initial inspection includes closure of the defect with repair of the atresia in 4–6 weeks or creation of a stoma. Rarely is the condition of the bowel amenable to primary anastomosis at the time of initial closure. If the atresia is distal, a stoma created at the umbilicus effectively decompresses the bowel and allows for early feeding. Proximal atresias may be adequately decompressed by nasogastric decompression.

## Suggested Reading

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