CHAPTER 16

Omphalocele

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INTRODUCTION

Exomphalus (also known as omphalocele) is a condition that is seen in newborn infants, and is thought to result from failure of the intestines to return to the abdomen after the migration into the umbilical cord that occurs between the sixth and tenth week after conception. The incidence of exomphalus has not changed over the last several decades. There is no known environmental, racial or geographic predilection, although in rare cases there may be a familial predisposition. Exomphalus is also associated with a lower than normal birth weight and gestational age.

Exomphalus is characterized by a central defect at the umbilical ring; a membrane composed of visceral peritoneum, Wharton's jelly and amnion covers the eviscerated abdominal contents. The umbilical cord inserts onto the exomphalus sac. The sac usually contains loops of small and large intestine, stomach and, in approximately 50% of cases, liver. The abdominal muscles are normally developed. Rupture of the sac is reported in 10–18% of cases. This can happen in utero, at time of delivery or after delivery.

Exomphalus is frequently associated with other anomalies, the most common of which are cardiac and gastrointestinal tract abnormalities. Chromosomal abnormalities are often seen, particularly in children with small defects that do not contain liver. Exomphalus is also associated with Beckwith-Wiedeman syndrome, cloacal exstrophy, and pentalogy of Cantrell.

- **Prenatal Diagnosis and Management.** Exomphalus is often suspected because of an elevated level of maternal serum α-fetoprotein. Prenatal diagnosis can be accurately made using prenatal sonography. Exomphalus can be differentiated from gastroschisis because of the location of the defect and the presence of a sac, although this may be more difficult if the sac has ruptured. If exomphalus is detected or suspected it is important to search for other abnormalities. In addition to a thorough ultrasound examination, amniocentesis for karyotype analysis should be recommended and fetal echocardiography should be done to look for major cardiac abnormalities. The mother should be transferred to a perinatal centre with experienced neonatal and surgical support. Although there is no clear evidence to support routine caesarean section, most practitioners will recommend caesarean delivery for fetuses with a large exomphalus to avoid liver injury and rupture of the sac.

- **Postnatal Management.** Immediate postnatal management consists of:
- Nasogastric tube placement to decompress the stomach
- Intubation, if the child is in respiratory distress
- Coverage of the sac with moist gauze and plastic foil
- Intravenous fluids
- Routine neonatal bloodwork
- Temperature control with a heating lamp
- Vitamin K administration
- Antibiotics, if the sac is ruptured

In addition, a thorough assessment for other abnormalities must be performed, which will directly affect decisions related to the care of the child. Detailed physical examination, radiological studies, echocardiography and abdominal ultrasound are important to identify any associated anomalies. Since some large defects are associated with pulmonary hypoplasia, careful assessment of oxygenation and ventilation should be done and respiratory support using intubation and mechanical ventilation should be instituted if necessary. The exomphalus itself should be evaluated to determine its size, contents, and integrity.

Newborns with abdominal wall defects require more intravenous fluids in the first few days of life than a normal infant, due to evaporative loss and third spacing. The daily intravenous fluid requirement must be adjusted based on the hourly urine output and other parameters for end-organ perfusion. Infants with a silo are at a particularly high risk of fluid, protein and temperature loss.

Based on the clinical status of the patient and the characteristics of the exomphalus, there are three broad categories of options for the surgical management of this condition:

1. Primary closure
2. Staged closure
   a. Skin
   b. Silo
   c. Sequential sac ligation
3. Nonoperative management with late closure

Figure 16.1, 16.2

Small to moderate defects, particularly those in whom the liver is not in the sac, may be closed primarily. The sac is removed.

Figure 16.3

The skin is undermined enough that a secure fascial closure can be accomplished. Absorbable or nonabsorbable sutures may be used. The skin is then closed. For very small defects, the umbilical cord can be left behind to give better cosmetic results.

Figure 16.1

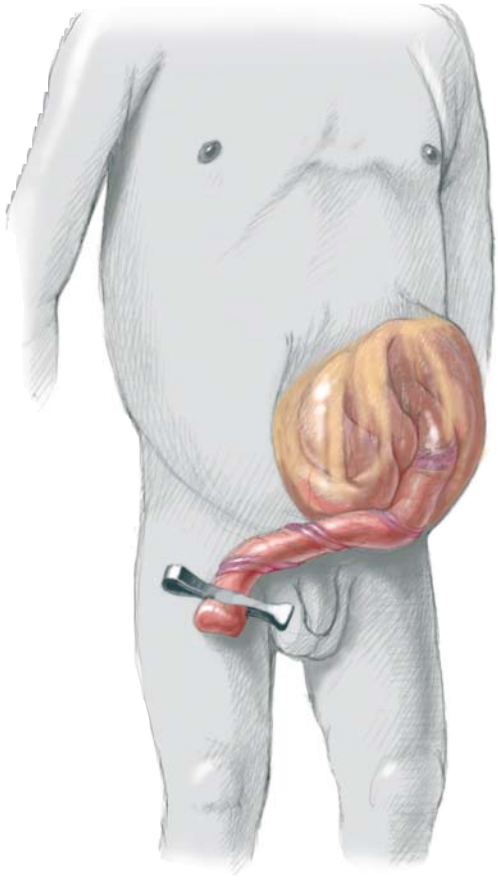


Figure 16.2

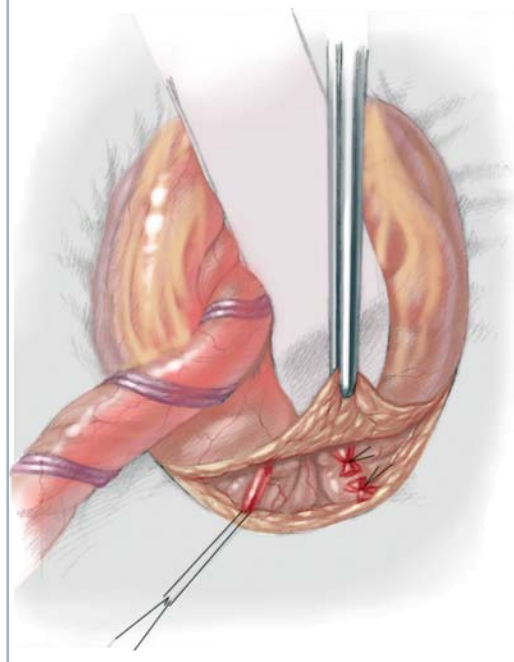


Figure 16.3

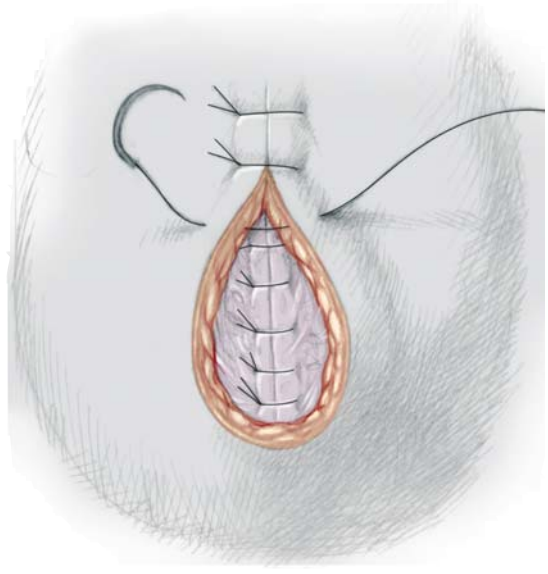


Figure 16.4–16.6

For larger defects, it may not be possible to close the fascia, but there may be enough skin to achieve skin closure over the viscera. This technique was originally described by Gross in 1948. The sac is usually removed, although some surgeons prefer to leave the sac intact and dissect between the edge of the sac and the skin to the level of the abdominal wall muscle. The skin is undermined as far out as possible, to permit skin closure with minimal tension. At this point, some surgeons opt to insert a patch into the fascia. The skin is then closed over the patch. In most cases of skin closure, the patient is left with a ventral hernia, which must be closed at a later time.

Figure 16.7

The use of a silo was first described by Schuster in 1967. The concept of a silo is to use a sheet of silastic reinforced with Dacron to gradually reduce the viscera over several days to a week, and then to definitively close the fascia and skin. This technique is useful for children with a large or ruptured exomphalus. The silastic sheeting is sutured to the edges of the musculae fascial layer, after as much of the intestine and liver as possible have been returned to the abdomen. Some surgeons also include the skin in these sutures. Although most surgeons remove the sac, some prefer to leave it intact and dissect between the edge of the sac and the skin to the level of the abdominal wall muscle. In some infants, the neck of the sac at the abdominal wall is relatively small, and the fascial opening must be enlarged to allow gradual reduction of the viscera.

Monofilament nonabsorbable sutures are then placed around the edges to avoid any gaps through which intestine can herniate. The silo is then closed over the top, by suturing it to itself. It should be perpendicular and suspended or supported to avoid any kinks in the intestine.

Figure 16.4

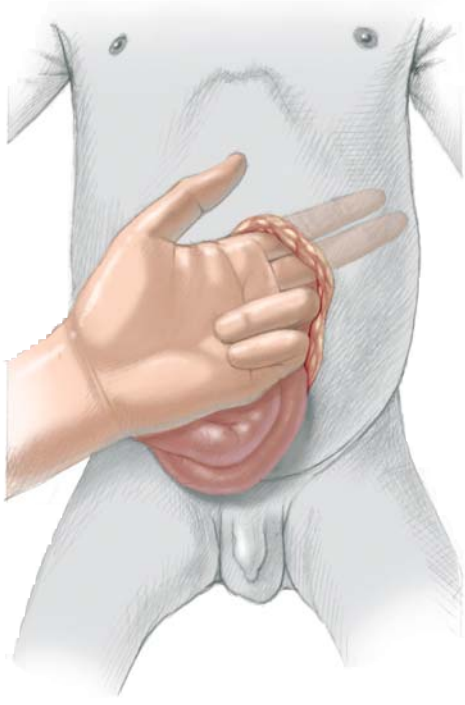


Figure 16.5

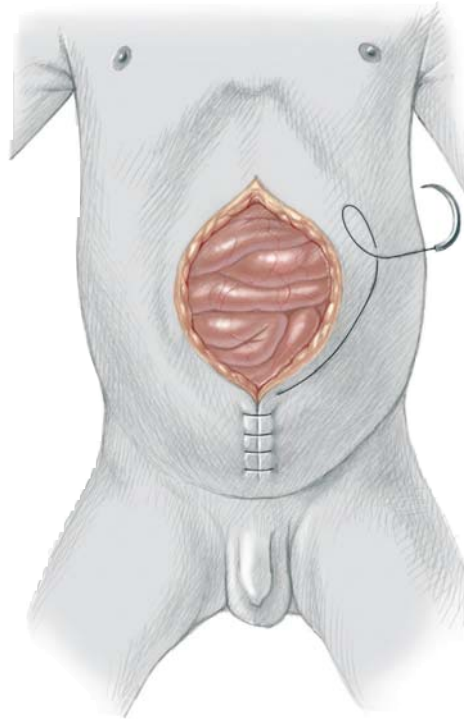


Figure 16.6

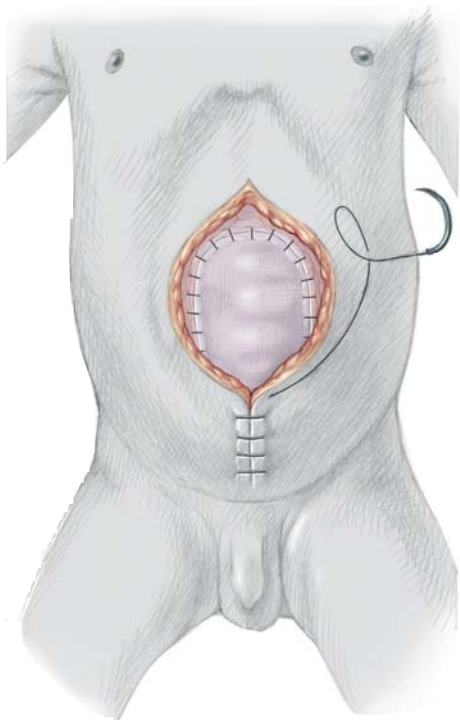


Figure 16.7

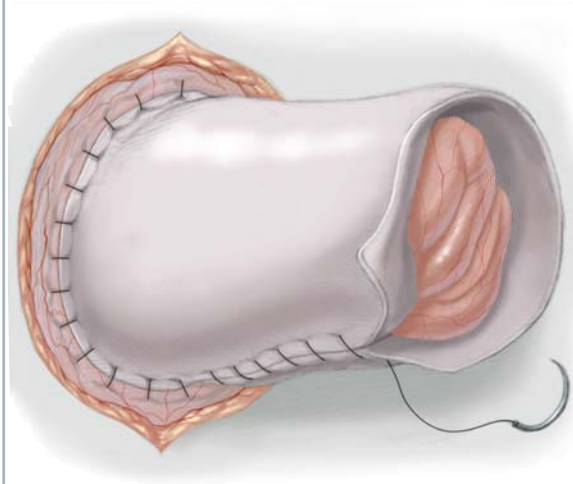


Figure 16.8

The sac is then gradually reduced at least once daily until all of the viscera have been reduced. Various techniques have been used to close the top of the silo, including sutures, umbilical cord clamps, umbilical tapes, and roller devices. Once the viscera are completely reduced, the child is brought back to the operating room usually after a week and the fascia and skin are closed.everal years in some cases.

Figure 16.9, 16.10

This recently described technique uses the exomphalus sac as a silo. It requires a sac which is relatively strong, and it is relatively difficult if the liver is adherent to a large part of the sac. However, it can be performed at the bedside in the nursery, with only minimal sedation. The technique involves gently kneading the sac to release minor adhesions between the sac and the intestine or liver. Traction is applied to the sac to slowly reduce the contents, and the sac is then twisted and ligated with umbilical ties. Once the viscera are reduced as much as possible, the child is taken to the operating room for definitive closure.

Some infants with exomphalus are very poor candidates for any kind of surgical intervention. This includes premature infants, those with chromosomal abnormalities, and those with significant congenital heart disease or pulmonary hypophasia. For these children it is best to cover the sac with a material which allows it to form granulation tissue and eventually epithelialize. Early on mercurochrome or iodine solution were used for this purpose, but there were problems with toxicity; this resulted in the abandonment of this practice. The use of plastic sheeting ("Op-site") has been described. We currently recommend silver sulfadiazine, which prevents infection and results in a good bed of granulation tissue. It takes several months for this to occur, and another several months for the granulation tissue to epithelialize. The resulting huge ventral hernia can be repaired electively whenever the child's underlying cardiac, pulmonary, or other conditions have improved. This may take several years in some cases.

Figure 16.8

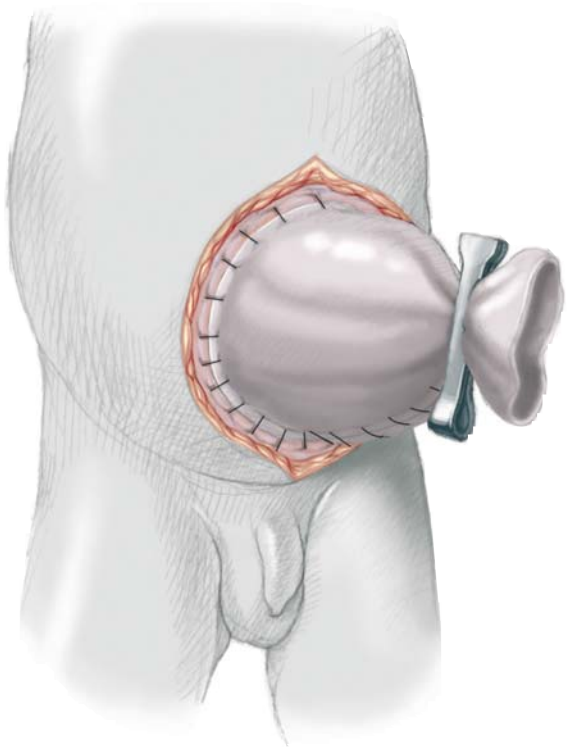


Figure 16.9

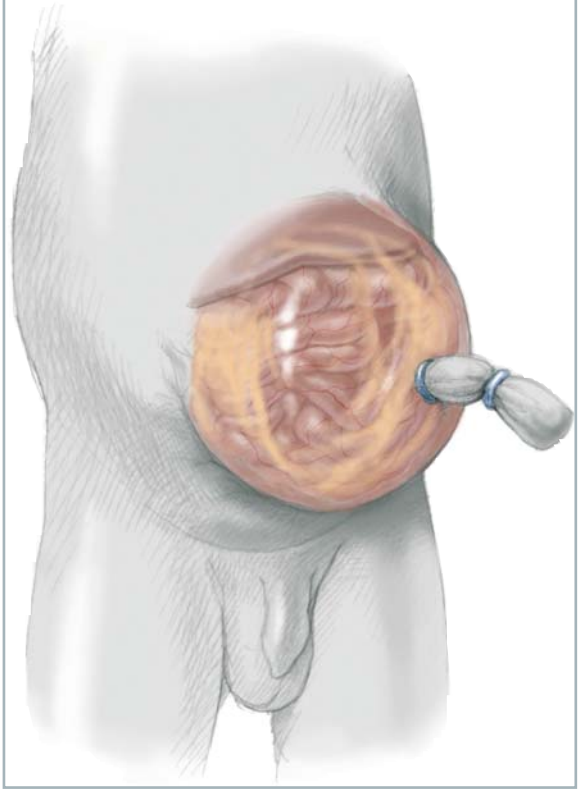
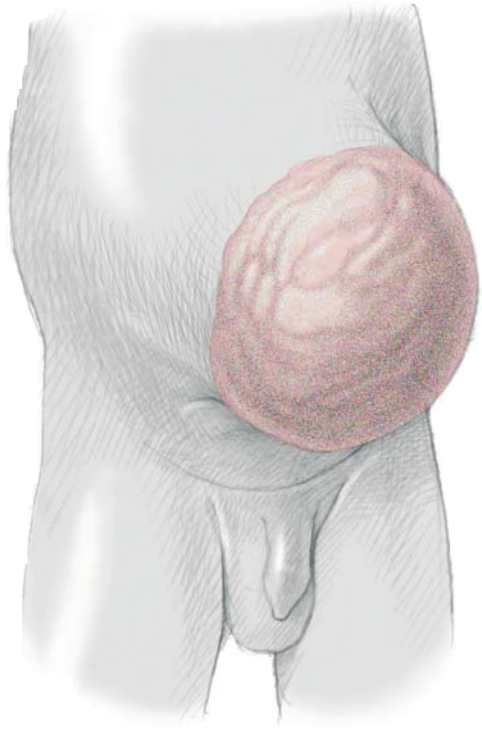


Figure 16.10



CONCLUSION

Care of the infant following definitive closure requires a neonatal intensive care unit for all but the smallest defects. Infants that have undergone repair of larger defects usually require postoperative mechanical ventilation for days to weeks, depending on their pulmonary status. It is important to carefully observe the child for signs of abdominal compartment syndrome, such as oliguria, acidosis, intestinal ischemia, and liver dysfunction. Infants with other congenital malformations require continued investigation and management as needed.

After closure of the abdomen, infants with exomphalus often develop an ileus, although intestinal function usually returns more quickly than seen in infants with gastroschisis. A nasogastric tube is therefore necessary initially. Total parenteral nutrition should be initiated early. Many surgeons place a central venous catheter at the time of the initial operation.

Intra-abdominal pressure monitoring using intragastric or intravesical catheters during closure can be an important adjunct to prevent abdominal compartment syndrome, which may result in high airway pressures, oliguria and intestinal ischemia due to decreased organ perfusion. Intra-abdominal pressures above 15 to 20 mmHg, or an increase in central venous pressure of more than 4 mmHg are associated with visceral ischaemia in both animal and human studies, and should stimulate consideration of conversion to a staged closure technique.

The outcome for infants with exomphalus is dependent on gestational age, the presence of associated chromosomal and structural anomalies, the presence or absence of pulmonary hypoplasia, and the size of the defect. Long-term problems that are commonly seen in these infants include gastro-oesophageal reflux, feeding disorders, and adhesive bowel obstruction. However, most of these issues can be corrected or improve on their own with time, and most infants with exomphalus who do not have severe additional anomalies or pulmonary hypoplasia do very well, and grow up to be normal individuals.

SELECTED BIBLIOGRAPHY

Bruch SW, Langer JC (2003) Omphalocele and gastroschisis. In: Puri P (ed) Newborn surgery. Arnold, London, pp 605–613

Grosfeld JL, Weber TR (1982) Congenital abdominal wall defects: gastroschisis and omphalocele. Curr Probl Surg 19:157–213

Hendrickson RJ, Patrick RJ, Janik JS (2003) Management of giant omphalocele in a premature low birth weight neonate utilizing a bedside sequential clamping technique without prosthesis. J Pediatr Surg 38:E14–E16

Hong AR, Sigalet DL, Guttman FM, Laberge JM, Croitoru DP (1994) Sequential sac ligation for giant omphalocele. J Pediatr Surg 29:413–415

Langer JC (2003) Abdominal wall defects. World J Surg 27: 117–124

Schuster SR (1967) A new method for staged repair of large omphaloceles. Surg Gynecol Obstet 125:837–850