

CHAPTER 56

CONGENITAL ANTERIOR ABDOMINAL WALL DEFECTS: EXOMPHALOS AND GASTROSCHISIS

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Introduction

Exomphalos and gastroschisis are the common forms of presentation of congenital abdominal wall defect.

Exomphalos (from the Greek *ex* = out; *omphalos* = umbilicus) refers to protrusion into the umbilicus. In its very mild form, a small loop of intestine protrudes into the base of the umbilicus; this is a hernia into the umbilical cord. In the more severe form, the defect allows protrusion of small intestine and other viscera, pushing the umbilical cord forward and distending its base into a cystic mass containing the viscera. This constitutes an omphalocele (from the Greek *omphalos*, *kele* = hernia, tumour). Omphalocele is more common, with a general incidence of 1:4,000 births. Omphalocele is a result of failure of formation and closing in of the anterior abdominal wall and could therefore be associated with other forms of impaired organ formation, which will determine the general prognosis.

Gastroschisis is a defect in the full anterior abdominal wall (from the Greek *gastro* = stomach—the term generally used for abdomen; *schisis* = fissure, tear, or gape) through which the abdominal content protrudes into the amniotic cavity.

Gastroschisis occurs in 1:10,000 births; although this is less common than exomphalos, in the Western world an increased incidence of tenfold is noted in young mothers with substance abuse. Gastroschisis is not due to or associated with impaired organ formation, but there could be complications from mass protrusion of viscera through a small defect, including vascular compromise, which in early foetal life could result in bowel atresia.

Demographics

The estimated birth prevalence of omphalocele in western countries is about 1 in 10,000 births while that of gastroschisis is about 2.5 in 10,000 births. The prevalence in sub-Saharan Africa is not known as there are no population based studies. While the birth prevalence of omphalocele has remained generally stable over the years, reports from industrialized countries (Europe, United States, Japan) indicate that the rate for gastroschisis is on the increase. When omphalocele is associated with other abnormalities, the aetiology is multifactorial and incidence varies with age of the mother. These abnormalities occur more in younger mothers; omphalocele alone is more prevalent in older mothers, however.

Aetiology/Pathophysiology

The aetiology of these conditions is not known. For omphalocele, the pathogenesis is related to the formation of the anterior abdominal wall and return of the midgut into the abdominal cavity. At the third week of gestation, three primitive divisions of the gut are identifiable as foregut, midgut, and hindgut. By formation of the folds, intraembryonic coelom becomes gradually separated from extraembryonic coelom. The fold initially consists of ectoderm and endoderm. The mesoderm later forms in between, and the folds close in on the umbilical cord and thus complete the anterior abdominal wall. Failure of mesoderm development results in defects. At the cranial portion, the defect could affect

the anterior wall of the chest (sternum, pericardium, and the heart, causing the classic features of the pentalogy of Cantrell). In the caudal aspect of the anterior abdominal wall, the defect may be associated with bladder exstrophy or varying degrees of anorectal anomalies. In the female, there may be a cloacal anomaly. Other anomalies that have been described as being associated include trisomy 13, 18, or 21 anomaly and the Beckwith-Weidemann syndrome.

The most common form is the central omphalocele, due to failure in the lateral folds. It may be classified in terms of shape, size, content, whether there are associated other anomalies, and whether the membrane coverage is intact or ruptured. More specifically:

1. Shape:

- Conical: includes hernia of the umbilical cord; usually small with broad skin edge diameter
- Globular: in which there is a large sac hanging on a relatively small diameter base and small abdominal cavity

2. Size of defect:

- Small diameter up to 5 cm, described as minor
- Diameter more than 5 cm, described as major

3. Content of the sac:

- Bowel loops only, small and large intestine sometimes on part of the stomach, bladder, and occasionally the ovary
- Bowel loops and liver

4. Associated with cardiac or other gross anomalies:

- Syndromic
- Nonsyndromic

5. Membrane coverage:

- Intact
- Ruptured membrane

For gastroschisis, a vascular accident of the right omphalomesenteric artery and abuse of vasoactive drugs have been implicated in the aetiology.

Clinical Presentation

Omphalocele (Figure 56.1) is an obvious abnormality in the newborn, presenting as a mass arising from a defect in the anterior abdominal wall covered by a membrane. The membrane is composed of an inner layer of peritoneum and an outer layer of amniotic membrane with Wharton's jelly between. It is attached by its base circumferentially to the skin of the anterior abdominal wall. The diameter of the base, the content of the sac, and the size relative to the size of the abdominal cavity will influence the decision for the method of management. Also important are whether the membrane is intact or not all around the circumference and whether the membrane or part of it is infected.

Other features to be examined are the possible associated congenital abnormalities. Such features as ectopia cordis, sternal defect, bladder

exstrophy, and anorectal anomaly are suggestive of a syndromic omphalocele. When loops of bowel are eviscerated, it is important to evaluate whether it is an omphalocele in which the membrane has ruptured or the diagnosis should be a gastroschisis.

In gastroschisis (Figure 56.2), the eviscerate bowel segment is commonly loops of small intestine and colon, sometimes stomach. The umbilicus is attached to the normal site in an intact anterior abdominal wall and the defect through which the bowel has herniated is usually in the right side of the umbilicus, separated from it by a small bridge of skin. There is no membrane over the bowel. A large segment of bowel with oedematous and congested bowel wall is seen in cases presenting late after birth. A bowel segment wholly covered by fibrinous material—not an obvious membrane as in omphalocele—suggests the gastroschisis had occurred early in utero. The fibrinous cover is a result of reaction to amniotic fluid.

Investigations

A normal, generally healthy-looking baby in whom the only obvious anomaly is the omphalocele needs immediate urgent exclusion of hypoglycaemia before routine investigation of haemoglobin and electrolyte checks are done. Other required investigations will be determined by evidence of associated anomalies. Abdominal ultrasound is used to ascertain the kidney, echocardiography is used if there are clinical signs of cardiac anomalies, and x-ray of the chest is used if there are signs relating to pulmonary anomalies. Chromosomal analysis may be required to rule out trisomy.

Gastroschisis requires a haemoglobin and electrolytes check as for preoperative preparation.

Treatment

Management of a baby with omphalocele or gastroschisis takes into consideration the following:

- treatment and care of the general state of the baby;
- specific treatment of the omphalocele; and
- management of associated anomalies.

General Care

The neonate should be well wrapped up in warm clothing to prevent hypothermia. Intravenous fluid should be commenced and the stomach should be kept decompressed with a nasogastric tube. The omphalocele, or the eviscerated bowel in the case of gastroschisis, should be well covered to reduce loss of heat and prevent injury. In intact omphalocele, when there are no signs of intestinal obstruction or anorectal anomaly, the baby can be commenced on oral fluids and feed. The baby should be transferred to a neonatal unit for further care.

Nutrition in Omphalocele and Gastroschisis

Small lesions may be expected to have no problem with normal feeds. When the baby vomits frequently in the early days of life, intestinal obstruction should be suspected. Commonly, it is due to ileus and bowel oedema. Atresia or malrotation are indications for early exploration. Babies with gastroschisis have poor bowel motility due to the long exposure of the bowel loops to the amniotic fluid in utero; thus, delay in enteral feeding is frequently experienced. It is advisable to commence parenteral nutrition where available and continue until the baby establishes normal gastrointestinal function.

Treatment of Omphalocele

The primary aim of treatment of omphalocele is to return the bowel into the abdominal cavity and close the anterior abdominal wall. The possibility to do so will depend on whether the viscera can be placed in the abdominal cavity without tension on the anterior abdominal wall, without intraabdominal compartment syndrome, and without pressure on the diaphragm, which would impair respiration.



Figure 56.1: Exomphalos major.



Figure 56.2: Gastroschisis.

Closure of Omphalocele Minor

Primary closure

Primary closure of the defect is possible in almost all cases of minor omphalocele. The membrane is cleaned and excised. The edges of the defect are determined and the fascial edges are closed, followed by skin closure.

Delayed primary closure

Primary closure may need to be delayed in minor omphalocele with an infected sac and oedematous abdominal wall. The sac is cleaned thoroughly, covered with Sofratulle® and gauze, and then wrapped with a soft crepe bandage. This is done daily, twice a day, morning and evening. If there is slough on the sac, the slough should be excised gently without causing bleeding. After 6 or 7 days, the omphalocele may be closed by excising the sac and closing the fascia and the skin. When closing the skin, an attempt should be made to construct a navel.

Closure of Omphalocele Major

Primary closure

The abdominal cavity is closed with or without excision of the sac. During primary closure, it is important to exclude intraabdominal compartment syndrome, which is determined by poor urine output, tight abdominal cavity, respiratory compromise due to splinting of the

diaphragm, and oedematous or dusky lower limbs due to poor venous return. The fascial layer and skin are closed separately, with possible construction of an umbilicus. Muscle flaps are sometimes created by making a release incision on the sides of the peritoneal cavity and mobilising the muscle medially to obtain fascial closure.

Staged abdominal wall closure

If closure of the fasciomuscular layer is not possible due to undue pressure on the diaphragm, only the skin may be undermined, stretched, and closed. This will heal, leaving a ventral hernia that can be closed at a later age.

Postoperatively, the child is monitored for adequate respiration and urine output. Oral feeds are commenced as soon as the child can tolerate them.

Skin stretching may need to be attempted to increase the surface area of the abdominal skin wall. This is usually possible and reduces respiratory stress. Skin flaps are sometimes created by making release incisions on the sides of the abdomen and mobilising the skin medially without attempting to appose the fascia and muscles.

Secondary abdominal wall closure

Secondary abdominal wall closure is repair of the ventral hernia by achieving fascial closure with native body wall and, where not possible, the use of a prosthetic material (Gor-Tex®, Surgisis®, Permacol™) followed by skin closure.

The general condition of the child must be good, with good nutritional status and haemoglobin of at least 10 gm/dl. General anaesthesia is used. The scar of the healed omphalocele is excised. The skin is undermined, the fascial edge identified, and the fascia mobilised and muscle edge identified. Any adherent viscera are released. The peritoneum is closed without tension. The fascia is then closed longitudinally with monofilament-interrupted sutures. The effect of the closure on respiration should be monitored by the anaesthetist. If there is respiratory compromise, the sutures should be released and the use of a prosthetic material, such as prolene mesh, Gor-Tex, Surgisis, or Permacol, should be considered, followed by skin closure.

Application of silo

Silo material is silicon or prolene or other nonirritant synthetic material that is nonporous and not adhesive. The mesh is constructed into a bag to fit firmly around the bowels and sutured tightly to the circumference of the fascia and subcutaneous tissues of the defect (Figure 56.3). Bogota bags or intravenous solution bags may be used as silos. Preformed silo bags are now available that can be placed on the omphalocele and applied firmly on the circumference; however, these are expensive. The baby is nursed in an incubator in supine position with the bag suspended from the roof of the incubator.

The baby is usually comfortable. Broad-spectrum antibiotics are administered. The circumference suture area is firmly packed and monitored for soaking, infection, or evidence of detaching. Over the days that follow, the bowel content gradually reduces into the abdominal cavity. The bag can get loose on the omphalocele. Further sutures or bands are then applied on the bag to keep it firm on the bowels; this may be required every 2 days. By 7 to 10 days, the omphalocele can be reduced sufficiently to enable closure. The most serious difficulties with silos are infection and detachment at the suture line.



Figure 56.3: Traditional silo bag.

Conservative Treatment

Conservative treatment involves nonoperative measures aimed at escharisation of the sac, which progressively contracts the scar, and encouraging rapid epithelisation from the edge. Various materials have been used, including mercurochrome solution, dilute silver nitrate solution, and 70–90% alcohol. The effect and complications on the baby have caused these solutions to be used less frequently. A useful method of conservative treatment is the application of closed dressing, which is applicable only for an intact sac. When the sac is ruptured, the silo is preferable.

For the dressing, the whole abdomen is cleaned with a plain antiseptic lotion and dried. A layer of Sofratulle® is laid to cover the whole sac. Two or three layers of soft cotton gauze are placed to cover the whole lesion. A soft crepe bandage, 4 or 6 inches, is applied around the circumference of the abdomen, thus maintaining uniform pressure on the omphalocele (Figure 56.4). The dressing is kept on for 24–48 hours and repeated with fresh materials. If the sac appears moist, the dressing should be done once every day. If there is evidence of infection, the dressing should be done twice a day.

By this method, the baby can be kept in the hospital for a shorter time than for other methods, usually 7–10 days, and can be discharged to continue further dressing on an outpatient basis.

By the time the omphalocele heals, there is a ventral abdominal hernia (Figure 56.5). This is repaired at a later stage, as described above.



Figure 56.4: Conservative management of omphalocele.



Figure 56.5: Ventral hernia formation.

Management of Gastroschisis

Gastroschisis has a better prognosis than omphalocele because the eviscerated bowel is usually a short loop of small intestine; the abdominal cavity would in most cases accommodate the herniated gut. There are usually no other serious associated congenital abnormalities.

Most gastroschisis can be repaired by primary closure. The small defect is extended, and exploration of the abdomen is done to exclude gut anomalies such as atresia or malrotation. The bowel is cleaned with warm saline and reduced into the abdominal cavity, and the wound is closed.

Gastroschisis is said to have occurred early in intrauterine life when the bowel has stayed out for long time before birth. The eviscerated loops are usually covered with fibrinous exudates due to amniotic fluid reaction. The abdominal cavity is usually small. Primary closure may not be possible. A silo is useful; the gut gradually reduces and the defect can then be closed.

Long delay before surgical intervention results in extrusion of more loops of bowel, which become oedematous and can be complicated by gangrene of the long segment of loop of gut. Resection of the gangrenous segment leaves a short length of small intestine.

Antenatal Diagnosis and Management

Omphalocele and gastroschisis can be detected early in pregnancy. Attempts to repair the defects intrauterine have not been successful. The main usefulness of antenatal detection is to transfer the baby in utero to a paediatric surgical centre and institute early treatment soon after birth. For large omphalocele, it may be preferred to deliver the baby by caesarean section to prevent rupture of the sac, which could occur during a normal vaginal delivery.

Key Summary Points

1. Omphalocele has a sac and has associated abnormalities that contribute to the mortality and morbidity of the condition.
2. Gastroschisis has no sac and has no associated abnormalities, except possible bowel atresia.
3. Primary repair has the best outcome; however, if this is not possible, consider other modalities to avoid intraabdominal compartment syndrome.
4. Hypoglycaemia must be excluded in omphalocele.

Suggested Reading

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