Exomphalos and gastroschisis

Rini Poddar MBBS DA FRCA
Lucy Hartley MB ChB FRCA

Exomphalos (omphalocoele) is a central abdominal wall defect that allows herniation of abdominal viscera into the umbilical cord. A membrane (consisting of Wharton’s jelly, peritoneum, and amnion) covers the viscera. During the 6th week of development, the embryonic intestine develops rapidly and migrates through the umbilical ring into the cord. In the normal fetus, the intestine returns to the abdominal cavity within the following 4 weeks. This return is associated with a 270° anti-clockwise rotation. Exomphalos occurs when the intestine fails to return to the abdominal cavity. It is postulated that this is due to delayed closure of the lateral folds in association with a large umbilical ring. Infants with exomphalos usually have an associated non-rotation or malrotation of their intestine. The liver, spleen, and ovaries are frequently present in the sac.1

Gastroschisis is a smaller defect in the abdominal wall, located to the right side of the anatomically normal umbilical cord. It is suggested that a vascular incident involving the omphalomesenteric artery is responsible for this anomaly.2 The complete intestine is often herniated through the defect; however, the testes, ovary, and liver are less commonly involved. There is no associated membranous covering present in gastroschisis. Therefore, the herniated bowel is directly exposed to the contents of the amniotic cavity. As a result, the bowel wall develops an inflammatory peel and the mesentery becomes thickened.

Exomphalos occurs in 1 in 13 000 births. The incidence of gastroschisis is between 1 in 6000 and 1 in 10 000 live births. Its incidence has steadily increased over the last 30 yr. There are definite associations between gastroschisis and low maternal age, low parity, maternal smoking, and the use of decongestants and aspirin. Male babies outnumber females by 3:2.3,4

Associated anomalies

Up to 72% of neonates born with exomphalos will have an associated congenital anomaly. Approximately 20% of these are cardiac in origin, most commonly tetralogy of Fallot or atrial septal defect.1 Other associated anomalies include:

(i) chromosomal trisomies—trisomies 13, 14, 15, 18, or 21;
(ii) Beckwith–Wiedemann syndrome—macroglossia, gigantism, and pancreatic islet cell hyperplasia;
(iii) pentalogy of Cantrell—exomphalos, diaphragmatic hernia, sternal defect, cardiac anomaly (commonly ventricular septal defect), and pericardial defect. This should be suspected if an infant with exomphalos presents with cyanosis.5
(iv) lower midline syndrome—bladder or cloacal exstrophy, imperforate anus, colonic atresia, vertebral anomalies, and meningomyelocele.

Gastroschisis is much less frequently associated with other anomalies. The majority involve the gastrointestinal tract; 10–15% will have an associated intestinal atresia. Meckel’s diverticulum and intestinal duplication have also been noted.1 Both abdominal wall defects are associated with prematurity and low birth weight for gestational age.3

Antenatal diagnosis and management

Both exomphalos and gastroschisis are routinely diagnosed by antenatal ultrasound. Once an exomphalos is identified, further investigations (including amniocentesis and fetal echocardiography) should be carried out to exclude other associated anomalies. Antenatal diagnosis allows timing, location, and mode of delivery to be planned in advance. Most neonates with exomphalos are allowed to deliver at term, but it is generally thought that neonates with gastroschisis benefit from early delivery (~37 weeks gestation) to limit bowel damage from exposure to amniotic fluid.

Some centres prefer to deliver neonates with a large exomphalos by elective Caesarean section to prevent damage to the exposed liver;
However, there is no evidence to support this mode of delivery in the smaller exomphalos or gastroschisis. Ideally, all infants with these defects should be delivered in a centre where both neonatal and surgical expertise are available.

**Post-delivery management and preoperative preparation**

After delivery, these neonates should be transferred to the neonatal unit for preoperative optimization. The mainstays of the initial post-delivery management consist of:

(i) fluid resuscitation;
(ii) care of herniated bowel/viscera and their blood supply;
(iii) bowel decompression using a nasogastric tube;
(iv) temperature regulation.

In neonates with exomphalos, the sac covering the bowel must be inspected to ensure that a rupture has not occurred. If the sac is intact, it should be protected with saline-soaked gauze. Neonates with ruptured exomphalos or gastroschisis are extremely susceptible to significant fluid and heat loss from the exposed bowel. To reduce these losses, the bowel is covered with a waterproof cellophane bowel bag. The neonate with gastroschisis should be nursed in the right lateral position with the bowel supported.

All neonates with an abdominal wall defect should be nursed in an incubator to reduce heat loss and a nasogastric tube should be inserted to facilitate bowel decompression. In addition to receiving maintenance fluid of 10% dextrose and 0.18% sodium chloride at 80 ml kg$^{-1}$ day$^{-1}$, they may require multiple fluid boluses to compensate for the excessive fluid losses mentioned above. These are usually given in 10 ml kg$^{-1}$ increments of 4.5% human albumin solution. Fluid requirement is determined by assessment of pulse, arterial pressure, and capillary refill. IV co-amoxiclav (30 mg kg$^{-1}$ 8 h$^{-1}$) is commenced routinely to combat infection.

Routine full blood count and capillary blood gases should be taken and abdominal and chest X-rays performed. In a neonate with exomphalos, an echocardiogram, kidney ultrasound, chromosomes, and head scan are also performed before operation to ascertain the presence of any associated anomaly. Cross-matched blood should be available for intraoperative transfusion if required.

**Surgical and anaesthetic management**

Surgery for exomphalos is undertaken electively unless a rupture of the sac has occurred, in which case, the defect is repaired within a few hours. A small defect may be repaired in a single stage, whereas larger defects can require the formation of a silastic pouch and gradual reduction. In extreme cases, the exomphalos is so large that the contents will not return to the abdominal cavity even with the formation of a pouch. In this scenario, the sac is left intact and allowed to slowly epithelialize. A ventral hernia results; this can be repaired at a much later date.

Surgery for repair of gastroschisis is usually performed within hours of birth unless bowel perfusion is compromised; in which case, the neonate is taken to the theatre as an emergency. There are a number of surgical options for the repair of gastroschisis. Traditionally, repair has been undertaken in the theatre environment. If possible, the exposed bowel is returned to the abdominal cavity in its entirety and a primary closure performed. However, some centres have moved towards a staged reduction of the herniated bowel using a silastic pouch, formed in theatre, with complete closure of the defect at a later date. In more recent years, a spring loaded, self-retaining silastic pouch has been developed which can be inserted on the neonatal unit without the need for general anaesthesia. This method also requires definitive closure at a later date. A further management option is elective delayed midgut reduction without anaesthesia; this was reported by Bianchi and Dickson in 1998 and Bianchi and colleagues. As this technique does not involve anaesthesia and remains controversial, it will not be discussed further.

All neonates are prone to hypothermia as a consequence of their large surface area to body weight ratio, reduced subcutaneous fat, and poor ability to shiver in response to cold. This tendency is exaggerated by the exposed bowel; therefore, the neonate should be transferred to the operating theatre in a warm incubator. Once in the theatre, active steps must be taken to reduce heat loss. These include the following:

(i) ensure theatre temperature $\geq 27$C;
(ii) place the neonate on a warming mattress;
(iii) heat and humidify inhaled gases to 36$^\circ$C.

Before induction of anaesthesia, pulse oximetry, electrocardiography, and non-invasive arterial pressure monitoring should be instituted, and the nasogastric tube aspirated to ensure bowel decompression. Invasive monitoring is not usually required. In the presence of a patent i.v. cannula, anaesthesia should be induced i.v. with thiopental 4 mg kg$^{-1}$; otherwise, it may be induced with sevoflurane in oxygen. In the neonate, muscle relaxation is produced using atracurium 0.5 mg kg$^{-1}$ or cisatracurium 0.15 mg kg$^{-1}$ and the neonate is intubated with an appropriately sized tracheal tube (usually 3.0–3.5 mm ID). A slight leak should be apparent with airway pressures of 20–25 cm H$_2$O in order to prevent compression damage to the tracheal mucosa. Correct tracheal tube placement should be confirmed by auscultation of the chest and end-tidal carbon dioxide measurement. A single-shot caudal epidural block may be performed to provide analgesia. The maximum dose of 0.25% 1-bupivacaine for this purpose is 0.8 ml kg$^{-1}$, which may be diluted to a volume of 1 ml kg$^{-1}$ with 0.9% normal saline.

Anaesthesia is maintained with oxygen, air, and a low concentration of sevoflurane (e.g. 1–2%). Nitrous oxide should be avoided in order to prevent further bowel distension. Muscle relaxation is maintained with a continuous infusion of cisatracurium 2 $\mu$g kg$^{-1}$ min$^{-1}$. Intraoperative analgesia may be provided by fentanyl 1–2 $\mu$g kg$^{-1}$ or morphine 25–50 $\mu$g kg$^{-1}$ if postoperative ventilation is anticipated. Maintenance fluid therapy with 10%
dextrose and 0.18% sodium chloride should be continued intraoperatively. Third space losses may be significant and require replacement with 10–20 ml kg$^{-1}$ boluses of crystalloids or colloid. Blood loss must be monitored throughout surgery and blood transfusion given if required.

The goal of surgery is to return the bowel to the abdomen and close the fascia in one operation. In some cases, the abdominal wall defect may need enlargement to enable this. The bowel must be inspected to identify any atretic segments or rotation abnormalities. If an atresia is found, but the bowel seems healthy, this can be repaired with a primary anastomosis. If not, a stoma is formed and reversed a few weeks later. Once the surgeon is confident that there are no other intestinal abnormalities, the bowel can be gradually returned to the abdominal cavity. Good communication between the surgeon and the anaesthetist is vital during this part of the operation as the intra-abdominal pressure may begin to increase causing respiratory compromise. A significant increase in peak airway pressure or decrease in tidal volume or oxygen saturation suggests that it may be impossible to achieve primary closure, and a silastic pouch will be formed instead.

Babies with extremely small defects may be extubated successfully at the end of the procedure. In these patients, reversal of residual neuromuscular blockade must be ensured by administering neostigmine 50 μg kg$^{-1}$ and glycopyrrolate 10 μg kg$^{-1}$ after a demonstration that spontaneous recovery of neuromuscular transmission has commenced. Neonates should be extubated only when they are fully awake, grimacing, moving vigorously, and breathing adequately.

**Postoperative care**

The great majority of babies who have undergone surgical repair of an abdominal wall defect will be returned to the neonatal unit intubated and ventilated. Sedation is usually provided with morphine 10–20 μg kg$^{-1}$ h$^{-1}$ and paralysis maintained with an atracurium or cisatracurium infusion at 0.5–1.0 mg kg$^{-1}$ h$^{-1}$ or 3 μg kg$^{-1}$ kg$^{-1}$ min. The duration of paralysis and sedation is governed by the ease and speed of return of the bowel to the abdominal cavity which may take up to 10 days.

Classically, the contents of the silastic pouch are reduced manually every 12–24 h. However, this is not always the case with the preformed, spring-loaded pouches. In this instance, the bowel is allowed to passively re-enter the abdominal cavity under gravity alone. During this period, and after primary repair, the neonate is monitored closely for signs of excessive intra-abdominal pressure. Apart from respiratory compromise, this may also cause reduced hepatic and renal blood flow, poor perfusion to the lower limbs, and intestinal ischaemia. If this ‘abdominal compartment syndrome’ is suspected, intra-abdominal pressure must be reduced. If a primary repair has been performed, the abdomen must be re-opened and a silastic pouch formed. If signs of excessive intra-abdominal pressure occur after reduction of the pouch contents, the tension on the pouch must be reduced and a proportion of bowel allowed to re-herniate. After complete return of the bowel to the abdomen, the neonate is taken to the theatre and the defect definitively repaired. Antibiotics should be continued until the silastic pouch is removed.

The first enteral feed is commenced once the volume of the nasogastric aspirate has reduced and become less bilious in nature. Full enteral feeding in neonates with gastrochisis is often not achieved for several weeks due to the damage to the bowel caused by exposure to amniotic fluid. As a consequence, total parenteral nutrition is commenced after operation, either via a surgical central line or via a percutaneous central line.

**Complications**

Complications are summarized in Table 1; they can be classified as gastrointestinal or non-gastrointestinal.

**Prognosis**

The mortality and morbidity associated with abdominal wall defects has decreased dramatically over recent decades due to advances in both surgical and neonatal care. In the 1960s, up to 70% of these neonates failed to survive. With improved preoperative and postoperative resuscitation, the outcome is excellent in 90% of cases. The vast majority of neonates without associated cardiac or respiratory abnormalities now survive to lead normal adult lives.

**References**


Please see multiple choice questions 9–12