Chapter

18 Neonatal Anaesthesia*

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Introduction

Some neonates may require specialist surgery during the first days or weeks of life. Many neonates presenting for repair of congenital abnormalities are born prematurely, which leads to additional challenges (discussed in Chapter 19). In this chapter, we consider common neonatal surgical conditions and their anaesthetic management.

General Considerations

Conditions requiring major general surgery in the neonatal period are:

- Oesophageal atresia (OA), with or without tracheoesophageal fistula (TOF)
- Congenital diaphragmatic hernia (CDH)
- Intestinal atresias
- Meconium ileus
- Malrotation and volvulus
- Hirschsprung's disease
- Imperforate anus
- Exomphalos and gastroschisis
- Necrotising enterocolitis (NEC)

Anaesthetic techniques should be tailored to the relevant surgical condition, but most of the key management principles are similar and require an in-depth knowledge of the anatomical, physiological and pharmacological differences particular to this vulnerable group of patients (discussed in Chapters 1 and 2). The following differences are of specific importance with respect to anaesthesia for neonatal general surgery.

Respiratory

- The intercostal muscles are poorly developed in neonates and contribute very little to the mechanics of respiration. Diaphragmatic breathing predominates and surgical conditions resulting in abdominal distension can significantly compromise ventilation.
- Oxygen consumption is twice that of the adult. Hypoxia develops very quickly if alveolar ventilation is compromised and may lead to profound bradycardia.
- All neonates are at risk of postoperative apnoea, and this is compounded by the presence of anaemia (Hb <100 g l^{-1}). Premature and ex-premature neonates are particularly susceptible; the incidence is decreased at 60 weeks postmenstrual age [PMA]).

Cardiovascular

- Many neonates with congenital defects presenting for surgery also have congenital heart disease (CHD). For some lesions, such as TOF and exomphalos, CHD may be present in 50% of cases. Cardiac lesions are often diagnosed antenatally. The presence of cyanosis should raise the possibility of undiagnosed CHD, although many cardiac defects are acyanotic. Neonatal surgery should be undertaken in specialist centres with preoperative echocardiography as standard practice. Surgery is not usually postponed in the presence of a cardiac lesion, but the cardiac physiology must be fully appreciated in order to ensure optimal management in the perioperative period (see Chapter 33).
- Occasionally, a prostaglandin infusion will be required to maintain patency of the ductus arteriosus. In lesions associated with very poor pulmonary blood flow such as severe tetralogy of Fallot, it may be necessary to perform a systemic to pulmonary artery shunt prior to general surgery. Close communication with the cardiologists is important to ascertain which lesion takes surgical priority.

* Many thanks to Dr Mark Thomas, who wrote the first edition of this book chapter, much of the content of which has been used in this revision.

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Thermoregulation

- Neonates have limited capacity to regulate body temperature, have poor thermal insulation and lose heat readily. Normothermia can be maintained by increasing ambient temperature, using warmed fluids and active warming strategies.
- The response to hypoxaemia is temperature dependent in neonates; under normal conditions peripheral chemoreceptors stimulate hyperventilation, but under hypothermic conditions hypoxaemia may result in hypoventilation and apnoeas.

Metabolic

- Neonates are at risk of hypoglycaemia in the first 48 hours of life, if they have low body weight (less than the third centile), if they have undergone prolonged surgery or if they have received extensive regional anaesthesia with a reduced stress response. Blood glucose should be monitored regularly, and glucose-containing fluids given to those at risk.

Haematological

- The haemoglobin level in neonates may be as high as 200 g l^{-1}, but it is predominantly fetal haemoglobin (60–90%) which confers relatively poor tissue oxygen delivery. Blood transfusion may be restricted to maintain Hb $>$ 90 g l^{-1} in a term neonate with no oxygen requirement, and $>$ 110 g l^{-1} if the neonate is aged under 1 week old, is intubated or has an oxygen requirement.
- Vitamin-K dependent clotting factors levels are very low at birth. Vitamin K (1 mg IM) should be given prior to surgery if it has not already been given at birth.
- Blood should be cross-matched for most surgery in the neonatal period. Coagulation may be disordered in neonates with bowel necrosis, and blood products are frequently necessary.

Neurological and Pain

- Nociceptive pathways develop within the first trimester and are particularly susceptible to maladaptive changes that may have long-term consequences for the neonate. Adequate analgesia must be provided.

Conduct of Anaesthesia

Preoperative Preparation

- **Fasting.** Breast-fed babies should be fasted for four hours prior to elective surgery, and babies receiving formula milk should be fasted for six hours. Clear fluids may be given up to one hour preoperatively.

Intraoperative Management

- **Airway.** The majority of neonatal surgical procedures require tracheal intubation and ventilation, as neonates have limited respiratory reserve and are prone to airway obstruction.
- **Induction.** Patients with small bowel obstruction are at risk of aspiration at induction. A modified rapid sequence induction should be performed, with gentle ventilation and the use of a non-depolarising muscle relaxant such as atracurium. It is reasonable to ask the parents to leave the anaesthetic room prior to induction so that full attention can be paid to the neonate at this critical time.
- **Volatiles.** The minimum alveolar concentration (MAC) for all inhalational agents is slightly greater in neonates than in adults. The neonatal myocardium is particularly sensitive to the depressant and vagotonic effects of inhalational agents. Sevoflurane is commonly used for induction followed by either sevoflurane or isoflurane for maintenance.
- **Analgesia.** A balanced anaesthesia technique using fentanyl will enable a reduced concentration of volatiles to be given, with the caveat that high-dose opioids will increase the requirement for postoperative respiratory support.
- **Neurotoxicity.** There has been concern in recent years that volatile agents may have a detrimental effect on the developing brain. Some anaesthetists advocate the use of spinal anaesthesia for surgical procedures such as inguinal hernia repair to avoid the potential disadvantages of general anaesthesia, but the

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surgery must be completed within one hour for this to be successful. The multicentre GAS trial investigating the effects of one hour of general anaesthesia with sevoflurane compared with awake regional anaesthesia for infants undergoing inguinal hernia repair showed no difference in neurodevelopmental outcome at five years. The rates of early postoperative apnoea were lower in the regional anaesthesia group.

- **Vascular access.** Ideally, two venous lines should be available and accessible during surgery (22G or 24G gauge). The anaesthetist should take care to preserve veins that may be used for long lines, particularly if use of parenteral nutrition is anticipated. For most major cases, an arterial line is extremely helpful, with distal arterial cannulation sites preferred. Complications associated with both arterial and venous access in neonates are common, and lines should be removed when they are no longer necessary.
- **Monitoring.** Standard monitoring should be applied and haematocrit, blood gases, lactate and blood glucose checked regularly.
- **Temperature regulation.** Maintaining normothermia is often a challenge during neonatal surgery. Fluid warmers and forced-air warming mattresses should be used and core temperature monitored.

Postoperative Analgesia

- **Paracetamol.** Comprehensive data analysis has been undertaken for the use of intravenous (IV) paracetamol in neonates, with specific dosing regimens suggested according to PMA. Intravenous paracetamol preparations have a concentration of 10 mg ml^{-1}, and great care should be taken to prevent accidental overdose due to confusion between the dose in ml and dose in mcg of the drug. There are reassuringly few reports of hepatotoxicity after overdose, particularly when the antidote, N-acetylcysteine (NAC) is administered promptly. Paracetamol should be prescribed regularly, not 'as required'.
- **Ibuprofen.** Ibuprofen is not licenced for use in infants under three months of age.
- **Opioids.** Intravenous morphine delivered via nurse-controlled analgesia (NCA) is used commonly after major surgery. Morphine increases the risk of postoperative apnoea in unventilated neonates; oxygen saturation and apnoea monitoring in a high-dependency unit (HDU) environment is essential. It is our practice to use morphine boluses of 10 mcg kg^{-1} with a 20 minute lockout and to avoid a background infusion. Oral morphine can be given at an effect-titrated dose of 0.05–0.1 mg kg^{-1} every four to six hours in carefully monitored term neonates. Extreme caution should be taken when giving oral morphine to non-ventilated premature infants due to the increased risk of adverse respiratory events.
- **Regional.** Epidurals are effective in neonates and may avoid the requirement for postoperative ventilation. Neonates have lower levels of plasma-binding proteins (in particular, alpha-1 glycoprotein), and higher free-plasma levels of local analgesic are possible. Our preference is to use plain 0.125% levobupivacaine, up to 0.3 ml kg^{-1} hr^{-1}. Local anaesthetic infiltration up to a maximum of 0.8 ml kg^{-1} of 0.25% levobupivacaine and NCA can be used as an alternative to an epidural.

Minimally Invasive Surgery (MIS)

Neonatal surgery is increasingly performed thoracoscopically or laparoscopically for procedures such as repair of TOF, congenital diaphragmatic hernia, duodenal atresia and malrotation. It is essential that the anaesthetist and surgeon work closely together to minimise complications during MIS. Anaesthetic considerations for MIS include:

- **Patient positioning and access.** The neonate may be positioned head up, head down or semi-prone and will be surrounded by equipment and monitors. There will be limited access to the tracheal tube (TT) and lines once the drapes are in place, so these must be checked and fixed securely prior to surgery.
- **Effects of pneumothorax/ pneumoperitoneum.** Specific complications from MIS in neonates include hypercarbia, desaturation, hypotension, metabolic acidosis and hypothermia and are seen more often during thoracoscopic procedures.
    - *Respiratory effects*: A cuffed or well-fitting uncuffed tracheal tube should be used to ensure that adequate tidal volumes are delivered during periods of reduced

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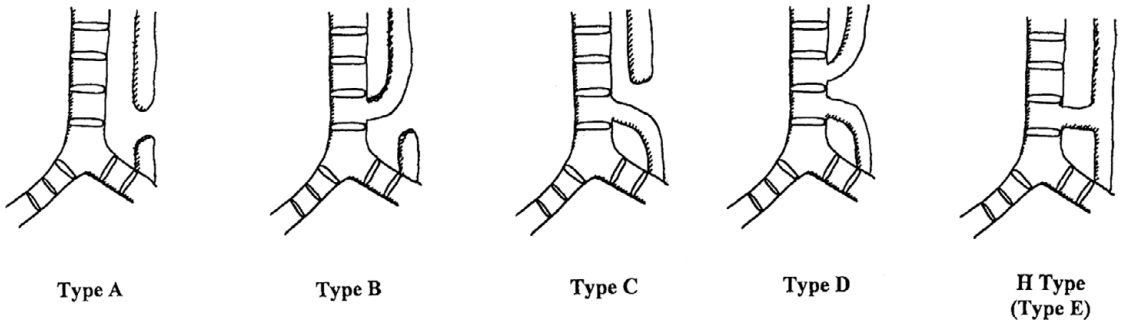

Figure 18.1 Gross's classification of oesophageal atresia and tracheal-oesophageal fistula.
Source: Gross RE. *The Surgery of Infancy and Childhood.* WB Saunders, 1954; figure by Thomas M.

respiratory compliance. The CO_2 insufflation pressure should be limited to 8 mm Hg. Systemic absorption of CO_2 can rapidly lead to high $PaCO_2$. Arterial blood sampling or transcutaneous CO_2 monitoring are invaluable for monitoring trends in these parameters. Minute ventilation will need to be increased to compensate for increased $PaCO_2$. It may be necessary to intermittently reduce the CO_2 insufflation to facilitate CO_2 clearance.

- *Cardiovascular effects*: Increased intra-abdominal pressure leads to reduced venous return, which may be particularly significant in neonates with CHD; isolated cardiac arrests have occurred during laparoscopic procedures. It is advisable to augment the preload with 10 ml kg^{-1} IV crystalloid before insufflation to preserve cardiovascular stability.
- *Thermoregulation*: The CO_2 insufflation gas is cold and may result in a reduction in body temperature.

- **Prolonged surgery.** Operating time should be limited to less than 100 minutes to minimise the adverse cardiorespiratory sequelae from insufflation.

Specific Neonatal Conditions

Oesophageal Atresia and Tracheoesophageal Fistula

Tracheal-oesophageal fistula (TOF) is a common congenital anomaly of the respiratory tract with an incidence of approximately 1:4500 live births. Oesophageal atresia (OA) and TOF are classified according to the anatomical configuration (Figure 18.1). The most common is the combination of a proximal oesophageal pouch with a distal TOF (Type C), which represents more than 80–85% of cases. Antenatal ultrasound may identify polyhydramnios, although many cases are not detected prenatally. Infants with OA become symptomatic immediately after birth, with excessive secretions that cause drooling, choking, respiratory distress and the inability to feed. Diagnosis is made by the failure to pass a nasogastric (NG) tube and a chest X-ray (CXR) which shows the NG tube coiled in the proximal oesophageal pouch (Figure 18.2). Patients with H-type fistulae may present early if the defect is large, but smaller defects may not be diagnosed until infancy or early childhood.

Additional clinical features relate to the presence of the VACTERL association (vertebral, anorectal, cardiac, tracheal-oesophageal fistula, renal and limb defects) or CHARGE syndrome (coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities and ear abnormalities). Congenital heart disease is seen in up to 40% of patients with TOF, most commonly ventricular septal defect (VSD) or tetralogy of Fallot. Echocardiography and renal ultrasonography should be performed in all infants in whom a TOF has been identified. The modified Spitz criteria can be used to identify risk factors for mortality, specifically birth weight <1.5 kg and the presence of major cardiac morbidity.

Neonates with TOF/OA require a Replogle suction catheter in the blind-ending oesophageal pouch preoperatively to help prevent pooling and overspill of secretions. This is a double-barrelled

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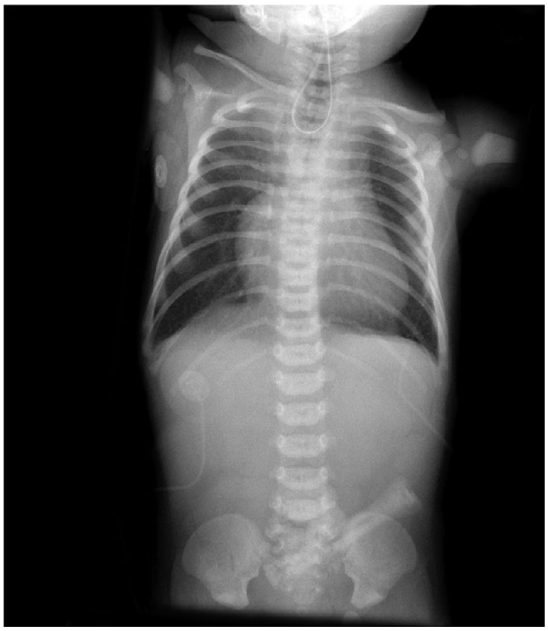

Figure 18.2 A coiled nasogastric tube suggestive of oesophageal atresia in a neonate with Down syndrome. *Source:* Thanks to Dr Frank Galliard, http://radiopaedia.org, reproduced with permission in the first edition.

oesophageal tube that is kept on continuous low-level suction. Inadvertent ventilation of the gastrointestinal tract may occur if the baby requires respiratory support preoperatively. This will result in bowel distension and further respiratory compromise. The requirement for preoperative ventilation is an indication for urgent surgery to close the fistula.

There is a role for microlaryngoscopy and bronchoscopy (MLB) prior to surgery to define the site and size of the fistula, exclude multiple fistulae and provide information to guide positioning of the TT. A recent multicentre audit in the United Kingdom showed that bronchoscopy was used in approximately 50% of cases. There is no standard of practice in anaesthesia or surgery in relation to the use of rigid or flexible bronchoscopy.

Anaesthesia

If an MLB is being performed, a gentle gas induction with sevoflurane in 100% oxygen is used, followed by topicalisation of the vocal cords with 1% lignocaine (2–3 mg kg^{-1}) when sufficient depth of anaesthesia is reached. An uncuffed TT is used as a nasopharyngeal airway and is passed into the posterior pharyngeal space, with anaesthesia maintained with 100% oxygen and sevoflurane. In our practice, the ENT surgeon then performs rigid bronchoscopy whilst spontaneous ventilation is maintained. Once the fistula has been identified, the surgeon or anaesthetist can advance the TT through the vocal cords into an appropriate position with the bevel of the TT facing anteriorly. Some centres advocate the use of a Fogarty catheter to block a large fistula, but this is not the usual practice at our institution. Other strategies include endobronchial intubation with withdrawal of the tube until bilateral air entry is obtained, keeping the bevel of the TT facing anteriorly. The fistula is usually in the posterior wall of the trachea, close to the carina, and this technique ensures that the tube is as distal as possible and less likely to result in ventilation of the fistula. Very rarely, it may be necessary to selectively intubate the bronchus in order to provide adequate ventilation for the procedure.

A non-depolarising neuromuscular relaxant should be given and the lungs ventilated with gentle manual positive pressure ventilation. Neonates with a large fistula may have problems due to gastric distension. If gastric distension does occur, disconnect the TT and apply gentle pressure to decompress the stomach via the TT. Whilst tempting, emergency surgical decompression via a gastrostomy will not be useful in this situation, as the gastrostomy offers a route of low resistance via the fistula, which will be ventilated in preference to the lungs. This is a particular concern in premature neonates with reduced lung compliance. Gastrostomy may, rarely, be useful to avoid life-threatening gastric rupture.

Surgery

Repair of the fistula can be performed using either an open or thoracoscopic technique. The main advantage of using a thoracoscopic approach is a reduction in the musculoskeletal sequelae. Suggested selection criteria include short-gap OA, birth weight $>$ 2 kg, cardiorespiratory stability with no major cardiac or pulmonary malformation and no more than one major malformation (such as duodenal or anal atresia). Some centres electively use high-frequency oscillatory ventilation (HFOV) to reduce hypercapnia and respiratory acidosis during thoracoscopic repair. This requires expertise in delivering HFOV in the theatre environment and a total intravenous

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anaesthetic (TIVA) technique to facilitate surgery and is not the current practice in our institution.

For open repair, surgical access is through a right thoracotomy (unless there is a right-sided aortic arch) with the baby in the left lateral position and their right arm above their head. The right lung is retracted. Gentle ventilation should be used until the fistula can be ligated. At the time of ligation, it is advisable to gently ventilate by hand to establish that it is not the right main bronchus or tracheal that is about to be tied off. Close cooperation with the surgeon is important if the patient is having frequent desaturations; intermittent reinflation of the compressed lung may be necessary. After successful ligation of the fistula, a primary oesophageal anastomosis is usually performed if the oesophageal gap is small. A transanastomotic tube (TAT) is gently placed with assistance from the surgeon; it must not be removed postoperatively and must be clearly labelled. In long-gap oesophageal atresia, a feeding gastrostomy is sited at the time of fistula ligation, and oesophageal anastomosis is delayed for a few weeks. The blind end of the oesophageal pouch is brought out into the neck to form an oesophagostomy to facilitate drainage of oral secretions.

Most babies will remain intubated, ventilated and sedated for one to two days postoperatively. Neonates who have an oesophageal anastomosis under tension are electively paralysed and ventilated for two to five days. It is better to err on the side of caution rather than risk disruption of the freshly sutured trachea should reintubation be necessary in the days immediately after surgery.

Infants commonly display a characteristic barking 'TOF cough' after TOF repair. Some will develop tracheomalacia, which may require aortopexy if symptoms are severe. Oesophageal strictures also occur, necessitating repeated oesophageal dilatations for dysphagia.

Congenital Diaphragmatic Hernia

Congenital diaphragmatic hernia (CDH) affects approximately 1:3,600 births and occurs when a defect in the diaphragm allows abdominal organs to protrude into the thoracic cavity. The exact aetiology is unclear, but the 'dual hit hypothesis' suggests that pulmonary hypoplasia is the primary disturbance, which then hinders the formation of the diaphragm. The resulting protrusion of abdominal contents into the chest further impairs lung development. Approximately 10% of patients have associated genetic or chromosomal abnormalities (trisomy 13, 18 and 21, Fryns syndrome, Cornelia de Lange syndrome, Beckwith–Weidemann syndrome or CHARGE syndrome) and a third of patients have one or more structural abnormality (cardiovascular abnormalities in 14% of patients). There are two types of CDH. Bochdalek hernias occur in 95% of cases, are posterolateral and almost always left-sided. Morgagni hernias are retro- or parasternal, usually smaller, predominantly right-sided and are associated with a better outcome.

Approximately 50% of cases of CDH are diagnosed antenatally. The absence of liver herniation is the most reliable predictor of survival. Cardiac defects have been shown to worsen outcome, and the presence of a small contralateral lung or a bilateral CDH are also poor prognostic signs. Some fetuses with a predicted poor outcome may benefit from the experimental intrauterine intervention of fetoscopic endoluminal tracheal occlusion (FETO). Its aim is to promote lung growth and consequently limit the developing pulmonary hypoplasia but is complicated by the risk of preterm delivery and concerns about tracheomegaly and tracheomalacia postnatally.

If not diagnosed antenatally, CDH usually presents with respiratory distress in the newborn, possibly in the presence of a scaphoid abdomen. A CXR characteristically shows bowel and sometimes other abdominal organs within the hemithorax, and the mediastinum may be displaced (see Figure 18.3). Resuscitation and medical stabilisation are required before definitive surgical repair. The majority of neonates will require tracheal intubation and ventilation immediately after delivery. The insertion of an NG tube may help to decompress the bowel and relieve pressure on the lungs. The pulmonary vasculature is usually underdeveloped, so pulmonary vascular resistance is high and pulmonary hypertension is common. Inhaled nitric oxide is often used, and IV sildenafil or prostacyclin may be required in severe refractory cases. Inotropes help to maintain organ perfusion and can decrease right to left shunting, thereby improving post ductal oxygen saturations. Neuromuscular blocking agents can improve synchronisation with respiratory support and optimise chest wall compliance when ventilation is challenging but should be avoided in the delivery room. Protective ventilation strategies should be

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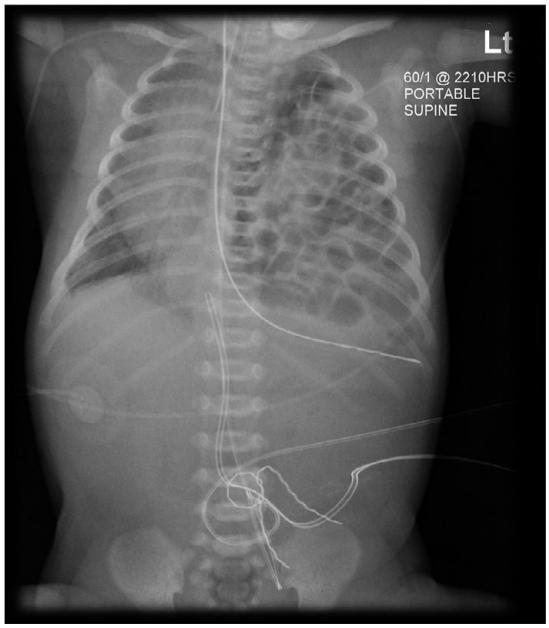

Figure 18.3 Left-sided congenital diaphragmatic hernia: the bowel can be seen herniating into the left hemithorax with displacement of the cardiac silhouette to the right.
Source: With thanks to Dr Frank Galliard, http://radiopaedia.org, reproduced with permission in the first edition.

used to avoid further lung injury, aiming to limit peak inspiratory pressures to 25 cmH_2O and allowing permissive hypercapnia. The VICI trial comparing HFOV to conventional mechanical ventilation found no significant difference in mortality when HFOV was used as an initial mode of ventilation. 'Gentle' mechanical ventilation is recommended as the initial ventilation strategy, with HFOV considered as rescue therapy if required. Extracorporeal membrane oxygenation (ECMO) may confer a survival advantage in the most severe cases of CDH, but its value in more moderate disease is less clear because of the high rates of ECMO-related complications.

The CDH EURO Consortium has published comprehensive consensus guidelines for postnatal management of infants with CDH. Elective surgical closure should be considered after clinical stabilisation once cardiorespiratory function is stable with clinical evidence that pulmonary hypertension is resolving. The CDH EURO consortium defines clinical stabilisation as normal blood pressure for gestational age, preductal SpO_2 85–95% with FiO_2 <0.5, lactate <3 mmol l^{-1} and normal urine output. Repair on ECMO is a viable treatment option in selected patients.

Surgery

Primary closure of a large defect may not be possible, and a patch is frequently needed to close the diaphragm. Open surgery is necessary for large hernias, particularly in the presence of liver herniation, and for sicker patients requiring higher ventilatory or cardiovascular support. Occasionally it may not be possible to close the abdominal wall without compromise to respiratory or cardiovascular stability and the risk of abdominal compartment syndrome. In this situation, the viscera can be covered and the patient left with a laparostomy, with definitive closure at a later date. Thoracoscopic repair has many theoretical advantages but is associated with more severe hypercapnia and acidosis, decreased cerebral oxygen saturations and longer surgery, the significance of which is unclear. Careful patient selection and multidisciplinary input is important in determining the appropriate surgical approach.

Anaesthesia

Periods of manual hyperventilation may be necessary to reduce hypercapnia during thoracoscopic repair, and conversion to an open procedure must be considered. Inhaled nitric oxide should always be available in theatre and started if a pulmonary hypertensive crisis is suspected. Simple interventions such as increasing FiO_2 and depth of anaesthesia, opioid administration and correction of acidosis are usually effective. Fluid overload should be avoided, as it may result in pulmonary congestion postoperatively. Inotropes may be required to optimise organ perfusion and manage intraoperative shunting. Pulmonary hypertension may persist in the postoperative period, potentially requiring further ECMO.

Intestinal Atresias

Congenital intestinal stenosis or atresia can occur at any point along the gastrointestinal tract. The neonate presents with bowel obstruction and specific clinical features depending on the level of obstruction.

Duodenal atresia occurs in approximately 1:6,000 live births. It has a male preponderance, and up to 45% of babies are born prematurely. Half of the patients have associated anomalies, particularly trisomy 21, malrotation and cardiac defects. Neonates present with bilious or non-bilious vomiting depending on whether the

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obstruction is distal or proximal to the ampulla of Vater. Abdominal X-ray demonstrates the characteristic 'double bubble' with an absence of gas distally. The patient should be resuscitated and stabilised, with intravenous replacement of fluid losses in addition to maintenance fluids, and an NG tube passed. Ultimate management is surgical. A laparotomy and duodenostomy are the primary procedures performed via a supraumbilical transverse or umbilical incision. There may be complete atresia or an intraluminal web that will need excising. The minimally invasive approach is feasible and increasingly being used. If the baby is otherwise well and stable, the aim should be to extubate at the end of surgery.

Jejunal/ileal atresia is a common cause of neonatal intestinal obstruction occurring in 1:3,000 live births. There may be a family history, and multiple atresias are not uncommon. Chromosomal abnormalities are much less common compared to duodenal atresia; however, patients with ileal atresia may also have cystic fibrosis. The atresia is thought to be caused by an intrauterine 'vascular accident' with interruption of the blood supply to affected segments. Neonates present with signs of intestinal obstruction and may have significant respiratory compromise necessitating respiratory support. Medical management includes NG decompression, IV resuscitation and maintenance fluids. Good outcomes can be obtained with excision and primary anastomosis at laparotomy. A proximal stoma and mucous fistula are formed as a temporising measure if the patient is unstable or the distal bowel is significantly compromised, and bowel continuity is restored at a later date. Postoperative ventilation may be required after prolonged surgery with significant fluid shifts. The primary cause of morbidity and mortality is short bowel syndrome or intestinal failure requiring parenteral nutrition, with associated risks of sepsis and liver disease.

Meconium Ileus

Meconium ileus is an intraluminal obstruction of the distal small bowel due to the presence of thick tenacious meconium. The majority are due to intestinal and pancreatic dysfunction secondary to cystic fibrosis. Once the obstruction has been treated, the infant should have genetic screening and a sweat test. Almost all such patients will require pancreatic enzyme replacement when feeds are introduced. The respiratory physicians should be involved early, although respiratory complications are uncommon in the neonatal period. Simple meconium ileus presents with distal intestinal obstruction. Abdominal X-ray demonstrates multiple dilated bowel loops with a 'soap bubble' appearance in the right-lower quadrant caused by mixing of air and meconium. Bowel perforation and meconium peritonitis may result in giant pseudocyst formation. Simple meconium ileus may be relieved with bowel washouts. Hypertonic enemas increase the risk of hypovolaemic shock due to sequestration of fluid in the gut. Surgery may be required if non-operative measures fail or in meconium ileus complicated by persistent bowel obstruction, perforation, atresia or volvulus. Surgical options include manual disimpaction via a proximal enterotomy or with the intraluminal injection of 4% N-acetylcysteine. Resection and anastomosis or stoma formation may be required for complicated cases.

Malrotation/Volvulus

Malrotation is congenital failure of the normal rotation and fixation of the bowel, resulting in a shortened mesenteric base and a tendency of the small bowel to twist around it (volvulus). This causes obstruction of the blood supply and lymphatic drainage to the small bowel in addition to luminal bowel obstruction, which can lead to ischaemia with necrosis of the entire midgut. It occurs in 1:6,000 live births but may be discovered incidentally. These neonates usually present with bilious vomiting, abdominal distension and pain. Fluid may be sequestered within the obstructed bowel, which can result in significant hypovolaemia and shock. Such neonates require resuscitation with large volumes of IV fluid and inotropic support. Systemic compromise and blood in the stools are later signs of volvulus in which significant ischaemia has already occurred. The gold standard for diagnosis is an upper GI contrast study which demonstrates the 'bird's beak' or 'corkscrew' appearance of duodenal obstruction. Malrotation with midgut volvulus constitutes a true surgical emergency, as the consequences are potentially catastrophic with loss of the entire small bowel.

The classic surgical approach is Ladd's procedure with derotation of the volvulus via laparotomy.

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Laparoscopy is increasingly being used, but the technique is still controversial, and early conversion to an open approach must occur if technical difficulties are encountered. The surgical options to address the ischaemic/necrotic bowel include excision of the necrotic segment, with or without anastomosis, or conservative management with a 'second look' laparotomy after 36–48 hours to determine whether perfusion has improved.

Anaesthetic considerations in the unstable neonate should focus on simultaneous resuscitation and expedited preparation and transfer to theatre. Coagulopathy is common in the presence of necrotic bowel, so blood and blood products must be available. The abdomen should be decompressed with an NG tube and respiratory support provided as needed. Invasive monitoring is very useful, and inotropic support may be required. Avoid deep inhalational anaesthesia in critically ill neonates. Derotation of the volvulus may lead to acute cardiovascular instability due to release of vasoactive compounds and lactic acid; intravenous calcium chloride and bicarbonate may be required to manage acidosis and hyperkalaemia. The infant who presents late with necrotic bowel may remain critically ill even after successful surgery and will require maximal support in NICU until perfusion is restored.

Hirschsprung's Disease

Hirschsprung's disease is due to aganglionosis, usually of distal bowel, and results in functional bowel obstruction. It is a multigenic disorder that is caused by a dysfunction in signalling pathways which are critical in the development of the enteric nervous system. Varying lengths of bowel are affected; short-segment disease affects the rectosigmoid region (80%), and long-segment disease extends proximal to the rectosigmoid region. Serial bowel biopsies are taken at the time of surgery to establish the line of demarcation between abnormal and normally innervated bowel. The most common presentation in neonates is bilious vomiting with failure to pass meconium in the first 24 hours of life. Signs of enterocolitis (fever, abdominal distension and diarrhoea) may also be present. This is a potentially fatal complication that must be identified early and managed aggressively to reduce the risk of sepsis, intestinal necrosis and perforation. Treatment is with fluid resuscitation and broad-spectrum antibiotics. Abdominal X-ray may show dilated bowel loops with an absence of air in the rectum. In the neonatal period, a suction rectal biopsy provides a definitive diagnosis when it fails to demonstrate ganglion cells.

Initial management aims to relieve the functional bowel obstruction, either with warm saline washouts or a defunctioning stoma into ganglionic bowel. A stoma is indicated if the neonate is unwell or has developed enterocolitis and perforation and has a grossly dilated colon or suspected long-segment disease. Definitive surgery consists of resection of the aganglionic segment, either after initial stoma formation or ideally as a primary procedure. Several 'pull-through' techniques have been described, usually performed when the infant is approximately three months of age or around 5–6 kg in weight. The timing and exact procedure undertaken varies between institutions and according to the length of abnormal bowel. The Duhamel procedure, in which the native rectum remains unchanged and a side-to-side anastomosis is stapled to the ganglionic bowel, is often the preferred technique for long-segment disease. Each technique can be performed open or with laparoscopic assisted intra-abdominal dissection and biopsies or entirely laparoscopically. For the laparoscopic approach, the patient is positioned in a steep Trendelenburg position. Blood loss is usually minimal. Caudal or epidural regional anaesthesia provides excellent perioperative analgesia after this surgery. Extubation at the end of surgery is usual.

Anorectal Anomalies

Anorectal anomalies occur in approximately 1:4,000 neonates and are diagnosed on clinical examination after birth. Imperforate anus with rectourethral fistula (males) or rectovesicular fistula (females) are the most common defects. Associated anomalies (genitourinary, spinosacral, cardiac, gastrointestinal and chromosomal) should be sought and further investigations (ECHO, abdominal X-ray, renal ultrasound) may be required. Initial management is supportive with IV fluids and gastric decompression with an NG tube. Surgical decompression and reconstruction are necessary. For 'low' anomalies, this involves a primary anoplasty with or without a diverting colostomy and a distal mucous fistula. For 'high' anomalies, a colostomy and a mucous fistula are formed, with later reconstructive surgery at one to

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two months of age. Several types of reconstructive surgery have been proposed, of which posterior sagittal anorectoplasty (PSARP) is the most common.

Stoma formation is considered relatively minor surgery in the neonatal period. Provided there are no significant sacral abnormalities, a caudal block provides excellent analgesia. Reconstructive surgery (PSARP) is usually performed in the prone position or as a combined abdominoperineal or laparoscopic-assisted procedure and may take several hours. Tracheal intubation is required, and IV access should ideally be placed in the upper limbs. Blood transfusion is rarely needed. The aim should be to extubate the baby at the end of surgery. Perioperative analgesia can be achieved with an epidural infusion or morphine NCA.

Cloacal Malformation

Cloacal anomaly is uncommon, occurring in 1:50,000 live births. It is seen exclusively in girls when the rectum, urethra and vagina fail to develop separately and instead drain via a single common channel onto the perineum. There is a high incidence of associated anomalies, so ECHO, renal and spinal ultrasounds are required. Initial management is supportive, with IV fluids, antibiotics, an NG tube to decompress the bowel and a urinary catheter to drain the urinary tract. When the baby is stable, a protective proximal transverse colostomy is performed to allow for adequate length for subsequent reconstruction. Endoscopy of the common channel is also undertaken to assess the internal anatomy and determine the length of the common channel. Definitive reconstruction (cloacal repair) to create three separate channels usually occurs at around two to three months of age.

Abdominal Wall Defects

Gastroschisis

Gastroschisis occurs in approximately 1:5,000 live births, is usually diagnosed on antenatal ultrasound and presents with herniation of bowel through a defect in the abdominal wall (usually right-sided), lateral to the umbilicus (Figure 18.4). The thickened bowel wall is covered in fibrin 'peel' due to exposure to amniotic fluid in utero. Gastroschisis is associated with prematurity, but the presence of other congenital abnormalities is unusual. The herniated bowel should be wrapped in cling film to minimise the large evaporative water and protein losses from the exposed bowel wall and to avoid damage to the bowel prior to surgery. Significant volumes of IV fluid may be required perioperatively.

Exomphalos

Exomphalos is rare, occurring in 1:10,000 live births. It is a midline periumbilical abdominal wall defect of variable size, covered by a membranous sac consisting of amnion (outer layer) and peritoneum (inner layer), with Wharton's jelly in between, and containing abdominal contents (Figure 18.5). The membraneous sac reduces evaporative losses and allows semi-urgent repair, unless

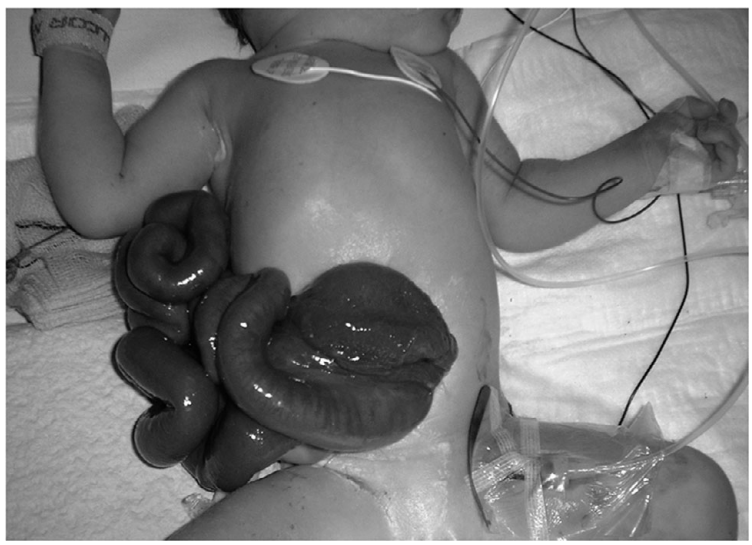

Figure 18.4 A neonate with gastroschisis.

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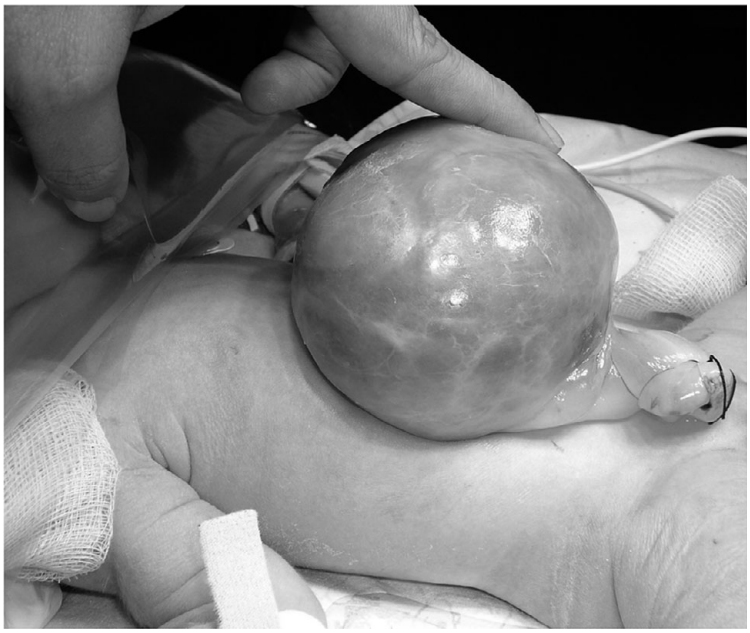

Figure 18.5 A neonate with exomphalos major.

the sac has ruptured. The defect may involve only minor herniation of the bowel (exomphalos minor) or may be large (> 4 cm diameter) and contain a herniated liver (exomphalos major). Giant exomphalos describes defects > 6 cm diameter. Associated congenital anomalies are common (> 50% of children) and include chromosomal abnormalities (trisomies 13, 18 and 21) and structural abnormalities, such as other midline defects and major cardiac defects. Beckwith–Wiedemann syndrome, an abnormality of chromosome 11 associated with gigantism, macroglossia, pancreatic islet cell hyperplasia with hyperinsulinism, organomegaly and hemihypertrophy, occurs in around 15% of cases. Exomphalos major is associated with pulmonary hypoplasia.

Surgery and Anaesthesia

The perioperative management for both of these abdominal wall defects is broadly similar. Initial management is supportive, with IV fluids, broad-spectrum antibiotics and an NG tube to decompress the stomach. Either a gas induction (avoiding nitrous oxide) or a gentle IV induction, followed by full muscle relaxation and ventilation, should be employed.

In exomphalos minor, the surgeon will attempt to reduce the herniated contents and close the defect in a single operation. In exomphalos major and gastroschisis, surgical closure in one stage may result in diaphragmatic dysfunction and compartment syndrome. Consequently, the bowel is placed in a temporary silo (prolene mesh sac), which is sutured to the abdominal wall. The anaesthetist and surgeon must work closely together to decide which option is the most preferable as determined by changes in lung compliance and gas exchange. Full muscle relaxation is essential at this stage. Some centres measure abdominal compartment pressure, for example with intravesical or intragastric pressure monitoring, to aid decision-making.

In the immediate postoperative period, these neonates should remain intubated and ventilated, particularly if primary closure has been performed. An opioid infusion for analgesia is usual. Many surgeons will request muscle relaxation for the first 24 hours after surgery. The silo is suspended above the baby to allow the bowel to be reduced under gravity, and a series of silo 'tucks' are performed on NICU over subsequent days (see Figure 18.6), facilitating early extubation. The baby then returns to theatre three to five days later for formal abdominal wall closure. Gut motility is commonly impaired after reduction of gastroschisis, and parenteral nutrition is commonly required.

Necrotising Enterocolitis

Necrotising enterocolitis (NEC) is predominantly a disease of prematurity and is covered in Chapter 19.

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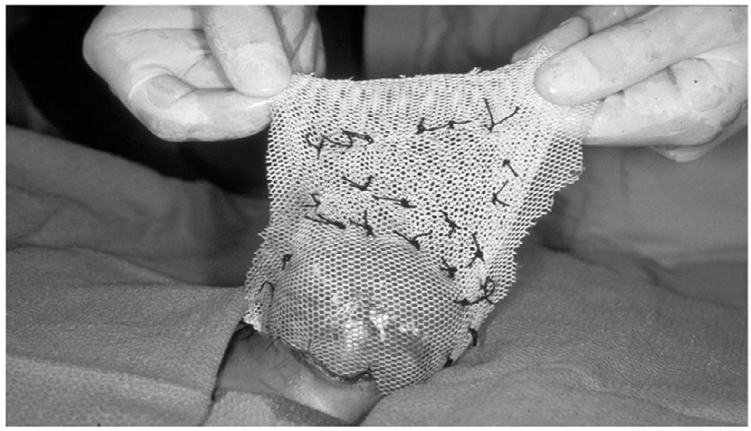

Figure 18.6 Management of gastroschisis with silo and sequential tucks.

Sacrococcygeal Teratoma

Sacrococcygeal teratoma (SCT) occurs in 1-2:40,000 live births. It is the most common neonatal tumour and is more frequently seen in females. Structurally, SCTs are classified as cystic (15%), solid (majority) or mixed. Perinatal mortality is around 30% and is more likely in rapidly growing vascular teratomas that act as arteriovenous malformations. These malformations lead to hydrops, high-output cardiac failure, preterm birth, and death. Caesarean section is indicated for large tumours to avoid rupture. At delivery, 90% of SCTs are benign, but 75% are malignant by one year of age if not resected. At birth, 90% are external midline sacral masses. Investigations should be performed to define the borders of the mass within the pelvis. Complications relate to vascularity, size and position of the teratomas.

Anaesthesia

Excision of SCT is high-risk surgery as it can haemorrhage acutely and dramatically. The perioperative risks relate to major haemorrhage from the highly vascular tumour in neonates who may be preterm and have pulmonary hypertension, high-output cardiac failure, coagulopathy and renal and hepatic impairment.

Once the airway is secured, arterial and central venous access is established, and the bladder is catheterised. The neonate is then turned prone, so vascular lines must be adequate and secure, as access to the baby will be limited. It is essential to plan for major blood loss, with cross-matched blood and blood products readily available. Intraoperative cardiac arrest has been reported from hyperkalaemia and hypocalcaemia secondary to massive blood transfusion. Hyperkalaemia has also been associated with surgical manipulation of a necrotic tumour. Blood must be transfused slowly, ideally via a peripheral cannula (not via a central line), particularly if the blood is not fresh; ideally fresh blood should be available. At the end of surgery, the neonate should be transferred to NICU in the prone position.

Surgery

The coccyx should be removed with the tumour for a complete excision. If the tumour has a small intrapelvic component, it should be resectable in the prone position. An abdominal approach may be indicated for larger intrapelvic tumours or in cases where early vascular control is required.

Key Points

- Neonates have specific physiological and pharmacological requirements.
- Congenital abnormalities requiring general surgery are often associated with cardiac and other anomalies.
- Neonatal anaesthesia is high risk and should be undertaken in specialist centres.
- Postoperative apnoea is common following neonatal surgery; many of these patients will require postoperative ventilatory support.
- Multimodal analgesia should be used for pain management.
- Vascular access can be challenging in neonates. Lines should be removed when they are no longer required.

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