# Congenital Diaphragmatic Hernias: Part 2

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## KEY POINTS

- Repair of a congenital diaphragmatic hernia (CDH) is not a surgical emergency. Haemodynamic and ventilatory stability must be achieved prior to surgical repair.
- Timing of surgery requires effective communication between the surgical, neonatal and critical care teams.
- Anaesthesia for CDH requires a thorough preoperative assessment, review of systems, an understanding of patient progress to date and an awareness of actual and potential perioperative patient specific concerns.
- Key intraoperative aims include ensuring adequate anaesthesia and analgesia, ensuring effective ventilation and avoidance of potential triggers of persistent pulmonary hypertension of the newborn (PPHN).
- Short-term postoperative complications that occur in the intensive care unit include bleeding, persistence of PPHN, pneumothorax, chylothorax, respiratory failure requiring oscillatory ventilation and sepsis.
- Long-term postoperative complications are multisystemic involving the respiratory, musculoskeletal, gastrointestinal and neurological systems.

## INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a rare defect of the diaphragm leading to herniation of the contents of the abdominal cavity into the thorax. While surgical correction is the definitive treatment, CDH repair is not a surgical emergency. Surgical repair should occur only in cases of haemodynamic and ventilatory stability (Table 1). Before any surgical intervention, patients must be resuscitated and stabilised in a paediatric critical care unit, which involves optimisation of oxygenation, ventilation and pulmonary hypertension (persistent pulmonary hypertension of the newborn; PPHN). Please refer to ATOTW 526 for further reading.

Anaesthesia for CDH is challenging. The type of defect, birth history, presence of associated congenital disorders and patient progress to date must all be carefully considered to determine the most suitable timing of surgery, surgical approach and a safe and effective anaesthetic plan.

## SURGICAL REPAIR

The objective of surgery is to reduce the herniated organs into the abdominal cavity followed by closure of the diaphragmatic defect. The traditional surgical approach is a transabdominal diaphragmatic herniorrhaphy. In

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| Clinical parameters |
| --- |
| $SPO_2$ >92% on $FiO_2$ < 0.5 |
| Mean arterial pressure >45 mm Hg (or normal for gestational age) |
| Weaning inotropes (noradrenaline and adrenaline <0.05 mcg/kg/min) |
| Lactate <3 mmol/L |
| Urine output >1 mL/kg/h |
| Pulmonary artery pressure <2/3 systemic pressure |
| Echocardiogram within 24 h of surgery with good right ventricular function |
| Weaning iNO doses <10 ppm |
| Hemoglobin >10 g/dL |
| Up-to-date chest radiograph |

**Table 1.** Clinical Parameters and Investigations That Determine Fitness for Surgical Repair of Congenital Diaphragmatic Hernia[1,2]

recent years, however, minimally invasive thoracoscopic repair has become the main surgical approach to CDH repair.[3]

## Open Repair

Open repair is mostly used for larger defects or in cases in which ventilation has been challenging or requires high-frequency oscillatory ventilation.[3] The incision for an open repair is subcostal on the side of the defect. Prosthetic patches are sometimes used when closing the abdomen in cases in which increases in abdominal and thoracic pressures cause haemodynamic and ventilatory instability.

## Thoracoscopic Repair

In less severe cases of CDH, thoracoscopic repair has become more common. The patient is positioned in the lateral decubitus position, and there is insufflation of the thoracic cavity with carbon dioxide to mobilise and reduce the abdominal organs.[4]

Originally, thoracoscopic repair was associated with longer operation times but with improvement in patient selection and surgical technique, this time difference is no longer seen.[5,6] Carbon dioxide insufflation can diffuse into the blood, creating a respiratory acidosis; this may be challenging to manage with changes in ventilation and is important because prolonged acidosis will worsen PPHN.

In either approach, once the organs are reduced, the diaphragmatic defect is then repaired either by way of primary closure or with a patch. Patch closure is commonly used in larger defects.[7]

# PREOPERATIVE ASSESSMENT AND REQUESTS

Careful preoperative assessment and preparation are key to the anaesthetic management of CDH. Assessment and preparation should include the following.

## Assessment

1. Antenatal history
2. Birth history
3. Presence of other congenital anomalies including cardiac, renal, neurologic and musculoskeletal
4. Cardiovascular status currently and trends since birth
   a. Dose and trend of inotropes, vasopressors and pulmonary vasodilators
   b. Serial echocardiogram results and evidence of improving pulmonary hypertension
   c. Haemoglobin and platelet values, clotting trends
   d. Mean arterial pressure
5. Ventilatory stability and trends since birth
   a. Size and length of endotracheal tube
   b. Presence of a difficult airway (intubation)
   c. Need for high-frequency oscillatory ventilation or extracorporeal membrane oxygenation
   d. Gas exchange on serial arterial blood gases
   e. Current ventilatory settings
   f. Pre- and postductal saturations
   g. Pneumothoraces, premature lungs

6. Fluid status
   a. Urine output
   b. Ongoing diuresis
   c. Glucose management
   d. Renal function and electrolytes
7. Sedation and muscle relaxation
   a. Current sedation
   b. Requirement for muscle relaxation to facilitate ventilation
8. Access
   a. Preductal arterial line
   b. Central access
   c. Intravenous (IV) canulae sites
   d. Ensure all access is working
9. Other investigations
   a. Result of renal ultrasound
   b. Result of cranial ultrasound

## Preoperative Requests

1. Blood products crossmatched and available
2. Haemoglobin >10 g/dL, clotting corrected
3. Echocardiogram within 24 hours before the operation
4. Chest radiograph
5. Drug infusions of inodilators/inotropes/sedation for transport

# INTRAOPERTIVE AIMS AND CONSIDERATIONS

## Communication

Communication between the anaesthetist and the surgeon must be clear and continuous throughout the procedure. The surgical team must be made aware of any haemodynamic and ventilatory instability as this may alter their operative plan. Similarly, operative progress, surgical concerns and unplanned surgical events should be communicated to the anaesthetist so that anaesthesia and postoperative care can be tailored accordingly. Instability particularly occurs on reduction of the herniated contents into the abdomen or during closure of the abdomen, so both teams should maintain extra vigilance during this time.[8]

## Ventilation

Ventilation strategies should be lung protective (Table 2). Pressure control ventilation is the most appropriate as it avoids high pressures and thus prevents barotrauma.[9,10] Excessive positive end-expiratory pressure should also be avoided. This is because overdistention of alveoli at the end of expiration compresses the pulmonary capillary networks. Compression of capillaries increases resistance to pulmonary blood flow, leading to worsening PPHN.

In determining a ventilation strategy, the preoperative critical care ventilation settings and the corresponding arterial carbon dioxide ($PaCO_2$) must be considered. Control of $CO_2$ is a key issue intraoperatively. High $PaCO_2$ will lead to a respiratory acidosis, which in turn causes pulmonary vasoconstriction and worsening PPHN. Regular intraoperative blood gas analysis is necessary to ensure close monitoring of $PaCO_2$.

The status of the lungs must also be considered. Abdominal viscera compressing the developing lungs leads to pulmonary hypoplasia of both lungs.[11] The lungs may also be premature, have suffered pneumothoraces, have poor compliance or have a high secretion burden. In the event of sudden instability, such as hypotension and hypoxia, a new pneumothorax should be considered. Pneumothoraces can occur intraoperatively to either lung, especially in cases of high ventilatory pressures.

| Intraoperative Ventilatory Aims |
| --- |
| 1. Lung-protective ventilation via pressure-control ventilation |
| 2. Low positive end-expiratory pressure |
| 3. Tight control of $PaCO_2$ |

**Table 2.** Intraoperative Ventilatory Aims for Paediatric Patients Undergoing Congenital Diaphragmatic Hernia Repair

## Haemodynamics and Pulmonary Hypertension

Also critical to anaesthesia for CDH is tight haemodynamic monitoring. The haemodynamic support established in critical care should be maintained in the operating theatre. Milrinone and any vasopressors or inotropes, if started, should all continue even at very low doses to allow quick titration in emergencies. Of note, milrinone is an inodilator that will reduce the workload of the right ventricle in PPHN.

If patients have pulmonary hypertension, due to PPHN, this causes increased right ventricular afterload, increased end diastolic volume and reduced output from the right heart. This can ultimately result in failure of the right ventricle and therefore must be monitored and treated aggressively intraoperatively. This involves avoiding potential triggers for pulmonary arterial vasoconstriction, such as avoiding hypoxaemia, hypercapnia, acidosis, hypothermia, pain and insufficient anaesthesia. Sildenafil and inhaled nitric oxide (iNO) from critical care should also be continued or be readily available in the event of haemodynamic instability due to right ventricular failure. This may require a specialised neonatal intensive care unit transport ventilator capable of delivering nitric oxide, which should be set up in theatre in advance prior to transfer of the patient to the operating theatre.

PPHN can be monitored by echocardiogram, which is identified by flattening of the interventricular septum, tricuspid regurgitation and a right-to-left or bidirectional shunt at the ductus arteriosus. Pressures in the right ventricle are estimated using the tricuspid regurgitation jet. Pulmonary artery pressures that are the same as systemic pressures are associated with a worse prognosis at any point in the perioperative period.[12]

Careful consideration should also be given to volume status. Blood loss monitoring, changes in haemodynamics and blood gas analysis will guide the necessity for blood products.

## Anaesthesia and Analgesia

Careful planning of transfer from the intensive care unit to the operating theatre is essential. Complications such as cardiovascular or respiratory arrest en route must be anticipated and prepared for. Emergency equipment and emergency drugs should be present.

Ongoing sedative infusions from critical care will reduce requirements at induction of anaesthesia. Anaesthesia for CDH can be achieved and maintained with a volatile agent such as sevoflurane or with total intravenous anaesthesia using propofol or other IV agents such as remifentanil. Favourably, sevoflurane, as with all volatile agents, has the protective advantage of reducing hypoxic pulmonary vasoconstriction.[13] It is, however, difficult to deliver volatile anaesthesia alongside iNO as iNO requires a high carrier gas flow technique to ensure no accumulation of iNO (measured in parts per million) within the breathing system.

Sevoflurane directly causes vascular smooth muscle relaxation,[14] leading to hypotension. Propofol also causes hypotension via vaso- and venodilation.[15,16] Excessive hypotension will reduce preload to the right heart, which is already under strain from the increased afterload caused by pulmonary hypertension. Vasopressors such as noradrenaline will aid in counteracting excessive hypotension and may need to be up-titrated, particularly after induction. Common intraoperative analgesia regimes for CDH include high-dose opioids, such as morphine, fentanyl or remifentanil by way of IV infusion or IV bolus. Muscle relaxation should be avoided as evidence suggests that it may be associated with reduced respiratory function with little added benefit.[2,13]

# POSTOPERATIVE MANAGEMENT

Postoperative care following CDH repair occurs in the intensive care unit (ICU). It generally involves the patient remaining intubated, ventilated, deeply sedated and adequately analgised with commencement of total parenteral nutrition in most cases.[9]

Several potential complications are worth keeping in mind in the hours and days after repair. Important short- and long-term complications are listed in Table 3. These include bleeding, persistence of PPHN, pneumothorax, chylothorax, respiratory failure requiring oscillatory ventilation and sepsis. Recurrence of the hernia is also possible.[9]

Close haemodynamic monitoring is essential postoperatively.[17] Postoperative bleeding may also occur. Bleeding is exacerbated by hypothermia and acidosis, both of which are associated with prolonged surgery as in CDH repair. Rewarming is essential postoperatively, as is the availability of blood products for effective and timely resuscitation when required.

Persistence or worsening of PPHN is another postoperative concern.[9] PPHN is aggravated by acidosis. Acidosis, in this instance, may occur as a result of hypercapnia secondary to difficult intraoperative and postoperative ventilation. It is also worsened by hypoxia and inadequate analgesia. Postoperative serial echocardiograms will help determine the severity and course of the PPHN. Echocardiograms will also determine the necessity for, and effectiveness of, treatment with inodilators or pulmonary vasodilators.

A chest radiograph is necessary following return to the ICU as postoperative respiratory complications are not uncommon. Position of the ETT and intravascular catheters should be confirmed. It is prudent to have a high index of suspicion for

| Short-Term Complications | Long-Term Complications |
|---|---|
| Respiratory:<br>• Pneumothorax<br>• Chylothorax<br>• Pleural effusion<br>• Atelectasis<br>Cardiovascular:<br>• Bleeding<br>• Persistence of PPHN<br>• Right ventricular failure secondary to PPHN<br>Infective:<br>• Respiratory infection<br>• Urinary infection<br>• Line infection<br>• Infected patch<br>• Infected wound<br>Surgery specific:<br>• Hernia recurrence | Respiratory<br>• Recurrent respiratory infections<br>• Chest wall abnormalities<br>Gastrointestinal<br>• Reflux disease<br>• Failure to thrive<br>Neurological<br>• Hearing loss<br>• Intellectual impairment<br>Musculoskeletal<br>• Lateral scoliosis |

**Table 3.** Short- and Long-Term complications After Congenital Diaphragmatic Hernia Repair. PPHN indicates persistent pulmonary hypertension of the newborn

pneumothoraxes. Most common are pneumothoraxes ipsilateral to the repair; however, contralateral pneumothoraxes may also occur in the context of aggressive or difficult ventilation. A pneumothorax usually fills with fluid unless it is a tension pneumothorax. A tension pneumothorax may be fatal and requires the urgent insertion of a chest drain.[18]

Overzealous crystalloid administration should also be avoided, especially in the context of PPHN and poor right ventricular function, as it may result in pulmonary oedema and pleural effusions. It is important to note that the space within the thorax that previously contained herniated contents will gradually be replaced with fluid in the days following repair. This is a normal process and should not be mistaken for a pleural effusion.

Postoperative sepsis requires early recognition and rapid initiation of treatment. A full septic screen must be carried out followed by initiation of empiric antimicrobial cover in the absence of a clear source or definitive microbiology. The most common sources include ventilatory acquired pneumonias, line sepsis, urinary tract infection and infection of the wound or surgical patch itself.[9]

Feeding, growth and nutrition are critical aspects of care following repair.[10] Feeding via total parenteral nutrition occurs in the days following repair as patients will be nil by mouth during this time. Guidance from dietetic colleagues will guide nutritional intake. Timing of commencement of enteral feeding differs depending on the status and progress of each individual patient and will be decided by the surgical team.

## LONG-TERM MANAGEMENT

The long-term sequelae of CDH are multisystemic (Table 3). These sequelae persist into later life and are a cause of significant morbidity. As such, follow-up and long-term management necessitate a multiple disciplinary approach.

Systematically, the complications are as follows[18]:

Respiratory:

1. Recurrent chest infections. These may occur up to 8 times per year. Recurrent infections are associated with patch closures, prolonged ventilation and HFOV.[11]
2. Chest wall abnormalities. Pectus excavatum, carinatum and chest asymmetry. These complications occur in cases involving the herniation of the liver into the thoracic cavity and cases involving thoracotomy.[2]
3. Chronic lung disease is most commonly caused by recurrent respiratory infections. Lung hypoplasia is also a cause of chronic lung disease.[11]

Gastrointestinal:

1. Failure to thrive.
2. Gastro-oesophageal reflux requiring fundoplication.

Musculoskeletal:

1. Lateral scoliosis. Thought to be secondary to ipsilateral lung hypoplasia. The type of repair (open versus thoracoscopic) does not appear to influence this outcome.[19]

Neurodevelopmental:

1. Hearing loss.
2. Intellectual impairment.

The pathophysiology of these neurodevelopmental sequelae is poorly understood.[20]

## SUMMARY


Congenital diaphragmatic hernias are rare congenital defects in the diaphragm leading to herniation of the abdominal contents into the thorax. Repair of CDH should be undertaken once there has been effective resuscitation and stabilisation in the ICU/critical care unit (CCU). This involves optimisation of oxygenation, ventilation and pulmonary hypertension.

Determining the most appropriate time to surgically repair a CDH is of the upmost importance. The decision to operate, as such, requires a multidisciplinary approach. Anaesthesia for CDH repair is complex and requires a thorough preoperative assessment, clear intraoperative communication with the surgeon, effective lung protective ventilation and avoidance of potential triggers of PPHN.

An awareness of the potential short-term complications that may occur in the ICU/CCU is essential in the hours, days and weeks after repair. Long-term sequelae are multisystemic and persistent into later life.


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