# CHAPTER 45
# Congenital Diaphragmatic Hernia and Diaphragmatic Eventration

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## Introduction

Congenital diaphragmatic hernia (CDH) is a group of conditions characterised by developmental defects in the diaphragm. The cause is disordered embryogenesis, resulting in incomplete fusion of elements giving rise to the diaphragm. CDH occurs at distinctive sites. The diagnosis can be made in the antenatal period, and can present in the early postnatal period with respiratory distress. Associated lung, vascular, and cardiac abnormalities lead to a high mortality (almost 50% overall), and prompt neonatal management is the most important influence on outcome. In this regard, surgical correction has become a nonurgent secondary intervention. Chromosomal abnormalities are found in 5–30% of cases (trisomy 18 and 13 are the most common). CDH can present outside the neonatal period in patients with minimal physiological compromise. The mortality is negligible in this naturally selected group.

## Demographics

The incidence of CDH is 1 in 2,500 to 1 in 3,500 live births. Left-sided CDH is more common than right-sided, with a ratio of 6:1. Bilateral lesions are reported, but they are invariably fatal. Ninety percent of CDH cases are found in a postero-lateral defect (Bochdaleck hernia), and 9% are found in an anterio-medial defect (Morgagni hernia). The remainder of cases comprise the relatively rarer forms of total absence of the diaphragm, absence of the central portion of the diaphragm, and oesophageal hiatal hernia. There is no gender or race predisposition.

## Aetiology/Pathophysiology

The diaphragm arises from four mesodermal elements in the embryo:

1. the pleuro-peritoneal membrane (fold);
2. the septum transversum (developing central tendon);
3. the dorsal mesentery of the oesophagus (crural precursor); and
4. somites of the body wall.

Fusion of these elements between the 5th to 8th week of intrauterine life separates the abdominal cavity from the thoracic cavity. The last element to close is the pleuroperitoneal membrane, the site of the Bochdaleck hernia, the commonest form of CDH. Return of the intestinal organs from the umbilicus around the 10th week of gestation can herniate into the chest if there is defective diaphragmatic development. Bowel loops within the chest compress the developing lung and cause lung hypoplasia (in both lungs, but in particular in the lung on the affected side). Development of type II alveolar cells that produce surfactant is also inhibited, resulting in relative surfactant deficiency.

Abnormal development of the pulmonary vasculature leads to pulmonary hypertension and increased pulmonary vasculature reactivity. Thus, the affected neonate is prone to episodes of hypoxia and hypercapnia, which in turn further increase the pulmonary hypertension and cause persistent foetal circulation. Persistent foetal circulation is a state of reduced pulmonary blood flow and pulmonary hypertension with severe right-to-left shunting through the patent ductus atreiosus and formanen ovale, which further worsens the hypercapnia and hypoxia. This vicious positive cycle can lead to severe physiological consequences in those most affected, and lung hypoplasia/pulmonary hypertension is the most detrimental pathophysiological process that affects outcome.

## Clinical Presentation

### History

#### *Antenatal*

In countries where routine antenatal ultrasound scanning is performed, approximately 50–85% of CDH are diagnosed on antenatal ultrasound scan. The features present antenatally are:

- polyhydramnios;
- absent stomach bubble or stomach bubble in chest;
- bowel loops in chest;
- mediastinal shift; and
- hydrops.

Foetal magnetic resonance imaging (MRI; see Figure 45.1) is also used in some centres for clarification of the diagnosis, to rule out associated anomalies, for planning, and for prognostic features. This is not widely practiced nor available.

Some features in the antenatal scan are associated with a poorer outcome. These are (1) hydrops, (2) contralateral lung-to-head circumference ratio <1.0, (3) diagnosis before 25 weeks gestation, and (4) associated cardiac abnormality. The role of antenatal contralateral lung-to-head circumference ratio in predicting outcome and indication

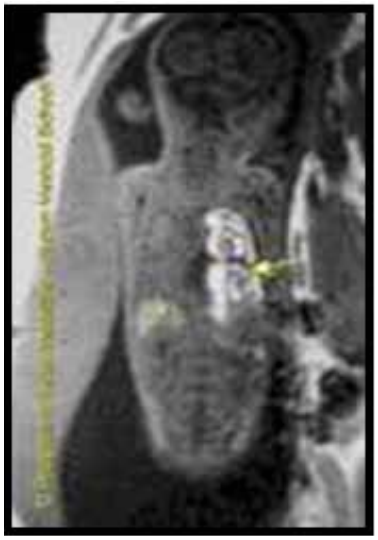

*Figure 45.1: Antenatal MRI scan showing a left CDH at a 32-week scan. Bowel loops are seen in the chest, and there is mediastinal shift.*

for foetal surgery is being questioned in recent studies, however.[1] Foetal surgery for CDH is being assessed in specialist centres, but as yet, there are no clear indications and benefit from this approach.[2,3]

***Postnatal***

Postnatally, the infant presents with respiratory distress. The timing of presentation is proportional to the degree of respiratory reserve; the later the presentation, the better the reserve and the baby's outcome. Grunting, tachypneoa, cyanosis, and poor feeding may be present.

### Physical examination

A general physical examination may reveal respiratory distress with grunting, use of accessory muscles and cyanosis. The affected hemithorax will have decreased respiratory movement. The trachea and apex beat may be deviated to the contralateral side. Diminished breath sounds with audible bowel sounds may be heard in the affected side. The abdomen is generally scaphoid in those presenting early. If presentation is delayed, however, this sign may not be present.

One particular presentation of CDH is with the constellation of the five malformations making up the pentalogy of Cantrell, a rare defect resulting from a severe mesodermal fusion failure:

1. Diaphragmatic hernia;
2. Lower sternal defect;
3. Pericardial defect;
4. Major cardiac anomaly; and
5. Epigastric exomphalos.

### Late and atypical presentations

In the absence of antenatal scanning and the absence of neonatal symptoms, some children may present later in childhood. They may present with poor feeding or vomiting and failure to thrive, poor respiratory reserve to strenuous exercise, or almost incidentally on an x-ray for a suspected chest infection. Subtle respiratory signs may be noted.

Cases are reported of children subject to minimal trauma, with severe respiratory symptoms, who undergo a chest x-ray and a diagnosis of tension pneumothorax is made (mistaking the herniated stomach for air in the pleural space). Needle or tube thoracocentesis of the chest is an avoidable iatrogenic complication if the x-ray is scrutinised carefully and the absence of a diaphragm noted, confirming a diaphragmatic hernia. Most of these cases are found to be a CDH at operation, although a traumatic rupture of the diaphragm is an alternative diagnosis.

### Differential diagnosis

The main differential diagnosis and the key features in differentiating them are:

- *Eventration of the diaphragm:* A thin rim of soft tissue shadowing may appear on the chest x-ray, suggesting that some diaphragmatic tissue is present. The diagnosis is best distinguished by using fluoroscopy to demonstrate paradoxical chest movement during respiration, but the distinction is sometimes made only at operation.
- *Congenital pulmonary airway malformations:* Congenital malformations of the airway and lung with cysts in the lower chest can mimic CDH on a plain x-ray of the chest. In these cases, however, the abdominal x-ray demonstrates a normal gas pattern with the nasogastric tube (NGT) in the abdomen, and a good diaphragmatic rim is usually seen. Usually no further imaging is needed to differentiate them, but a computed tomography (CT) scan is helpful in difficult cases.

## Investigations

A plain anterior-posterior radiograph of the chest is diagnostic in most cases. The x-ray should be combined with a plain abdominal x-ray with a nasogastric tube in place. Features of the common Bochdaleck hernia on the radiograph are (see Figures 45.2–45.4):

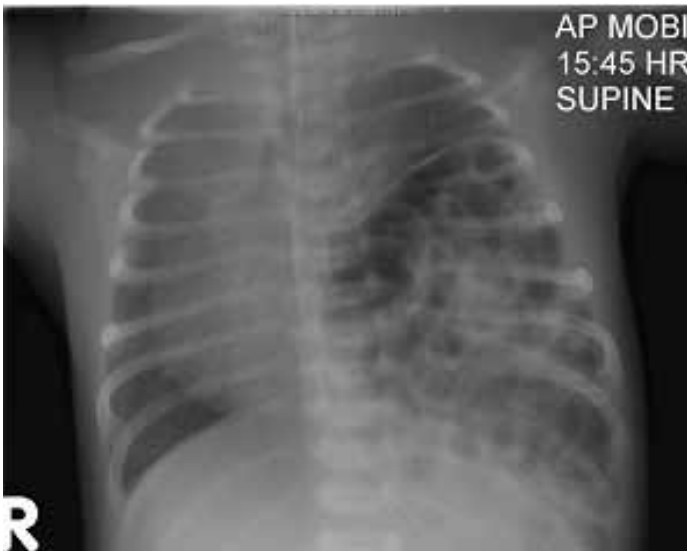


*Figure 45.2: Chest x-ray showing a left CDH. Bowel loops are seen in the chest, and there is mediastinal shift. The appearances could be similar to congenital lung cysts, and an abdominal x-ray is needed to confirm the diagnosis.*

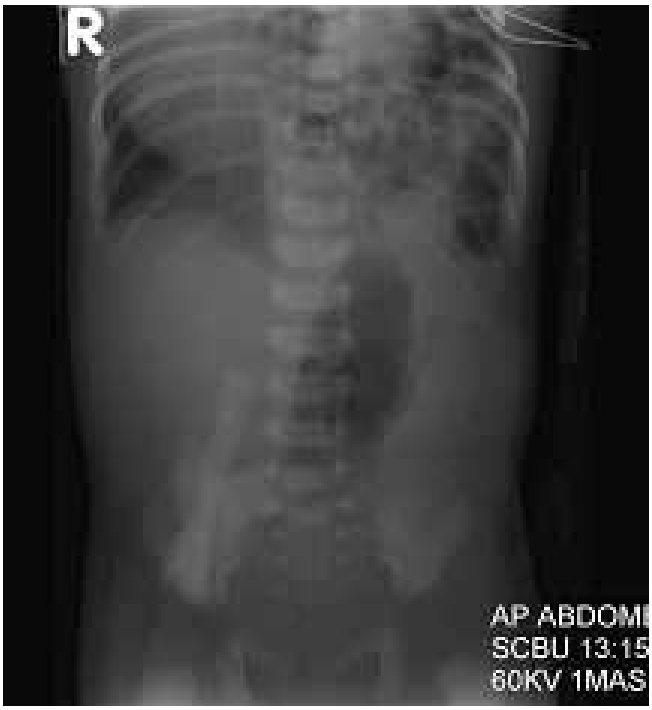


*Figure 45.3: Abdominal x-ray of the same patient as in Figure 45.2. Bowel loops are seen in the chest, and there is paucity of gas in the abdomen, confirming herniation. In this patient, the stomach did not herniate; therefore, the NGT is in the abdomen.*

- absence of the diaphragm;
- bowel loops seen in the chest, with paucity of loops in the abdomen;
- tip of NGT in the chest (only if stomach is herniated);
- mediastinal shift; or
- with right-sided lesions, a radio-opaque lesion replaces the lung tissue.

With a Morgagni hernia (see Figure 45.5) the features include a radiolucent shadow overlying the heart. A lateral view is helpful in showing this to be in the anterior mediastinum.

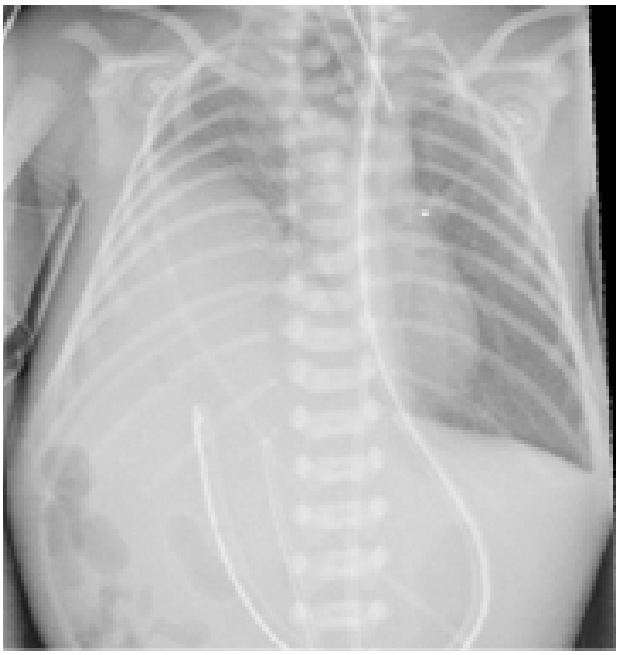

*Figure 45.4: Chest and abdominal x-ray of patient showing a right-sided CDH. A soft tissue shadow is seen in the chest and represents the herniated liver. Also note in this patient the presence of a dilated trachea (dilated radiolucent pouch in lower neck/upper chest) as a result of antenatal tracheal occlusion. Remnants of the balloon used is seen as a radio-dense dot in the left side of the chest.*

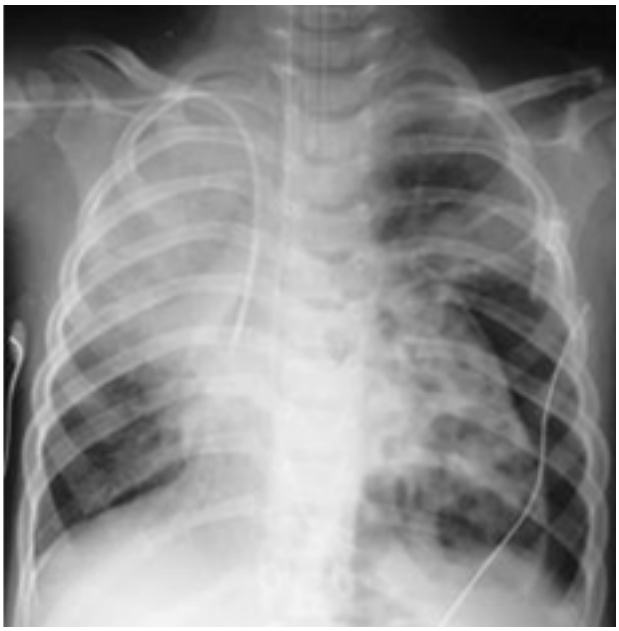

*Figure 45.5: Chest X-ray of patient showing a left-sided Morgagni hernia. The bowel loops are seen overlying the cardiac shadow, and loops can be traced from the abdomen just to the left of the midline*

Other investigations and their indications are:

- *An echocardiogram* is useful in evaluating cardiac function (shunting, ejection fraction, cardiac output, and changes with inotropic support) and in outlining any cardiac anomalies associated with CDH (atrial and ventricular septal defects).
- *A contrast study* may be indicated in cases of suspected Bochdaleck hernia that do not have a definitive diagnosis on plain radiograph. A contrast enema or meal with follow through may delineate bowel contents in the chest.
- *A renal ultrasound scan* is useful in ruling out any renal abnormalities.
- *Fluoroscopy* of the diaphragm may help differentiate between eventration and CDH in cases that are difficult to distinguish. *Ultrasound* of the diaphragm is less sensitive in picking up paradoxical movement than fluoroscopy, and is sometimes false negative.

## Management

The most important management is the resuscitation and stabilisation of the newborn by an experienced neonatologist. If the diagnosis is suspected, avoid bag and mask positive-pressure ventilation (to avoid intestinal distention and possible worsening of respiratory compromise).

The management of CDH is a complex one that involves specialist neonatal ventilatory and cardiovascular support in severe cases.[4,5] The essence of neonatal management can be summarised as follows:

- Prompt endotracheal intubation in the delivery room for respiratory distress.
- Replogle tube or wide-bore nasogastric tube insertion.
- Chest and abdominal x-ray to confirm diagnosis, assess NGT position, and exclude other diagnoses.
- Early measurement of blood gases, repeated at regular intervals to aid management.
- Surfactant administration, used in some centres in selected cases.
- Preductal oxygen saturations maintained at 85–90%.
- Minimal ventilation pressures to reduce barotrauma (iatrogenic injury from ventilation strategies may be significant and should be minimised).
- Volume resuscitation and vasopressors (dopamine and dobutamine) often required to maintain systemic blood pressure (BP) and reduce right-to-left shunting.
- Pulmonary vasodilatation with inhaled nitric oxide and occasionally other vasodilators (e.g., nitroprusside).
- Consideration of high-frequency oscillatory ventilation when conventional ventilation fails or when peak airway pressures remain high (>30 cm $H_2O$).
- Extracorporeal membrane oxygenation (ECMO), which has not offered consistent beneficial results in most studies. Oxygenation index ($FiO_2$ × mean airway pressure × 100/$PaO_2$) can be used to predict the need for ECMO in centres where this is offered. An oxygenation index value of >40 is an indication of severe respiratory failure and the need for ECMO.
- Initially not feeding by mouth (but trophic feeding is not strictly contraindicated in those stabilising with no signs of obstruction). Reliable central venous access is required for administration of drugs or fluids and/or parenteral nutrition.

One method used to predict outcome in the postnatal period is a formula developed at the Red Cross Hospital in South Africa,[6] (respiratory rate × $PCO_2$ × $FiO_2$ × mean airway pressure/$PaO_2$ × 6000), based on the first arterial blood gas obtained on initiation of resuscitation. A value greater than 5 was used as a cutoff between survivors and nonsurvivors, with 16/16 (100%) of patients above this value dying and 17/20 (85%) below this value surviving. Overall, it had a 91% predictive value.

## Surgery

Surgery is usually contemplated only in those who stabilise and improve on medical management. Stability is indicated by a decreased ventilatory requirement (transition from high frequency to conventional ventilation being a good sign of improvement), decreased oxygen requirements, return of haemodynamic stability, and weaning

off inotropes and pulmonary vasodilators if they were required. In those patients who have little cardiorespiratory compromise, a period of 24 hours (the so-called "honeymoon period" in CDH) to allow any instability to announce itself is prudent. Although there may be no long-term advantage of early versus delayed surgery,[7] a somewhat delayed approach (24 to 48 hours) may allow patients with significant cardiopulmonary disease, who would not survive despite any operative intervention, to be selected. In an otherwise stable patient, however, any long delay can be detrimental.[8]

The infant is taken to the operating theatre, and antibiotics, if not already administered, are given at induction. The operative steps are summarised as follows:

1. A transverse supra-umbilical incision is made.

2. Rarely, for large right-sided lesions with a larger proportion of the liver in the chest, a thoracoabdominal incision is required.

3. The intestines or viscera are inspected and gently and gradually reduced from the chest. Often, on the left, the spleen is particularly difficult to reduce without causing injury, and a finger or retractor introduced into the chest can be use to guide it into the abdomen. Rarely, the defect in the diaphragm needs to be enlarged to facilitate this. In right-sided lesions, reduction of the liver can be associated with altered venous return to the heart due to reconfiguration of the inferior vena cava; this should be anticipated and communicated to the anaesthetist.

4. After reduction of the abdominal contents, the chest is examined for a hernia sac. This is best done by grasping and incising over a lower rib to free a sac if present. The entire sac should be excised. The hypoplastic lung can then usually be seen in the chest.

5. The defect in the diaphragm is inspected, and the decision for primary or reinforced closure is assessed. Mobilisation of the leafs of muscle, in particular on the posterio-lateral aspect, can increase the amount of muscle available.

6. Primary closure is then achieved with interrupted nonabsorbable sutures. With large defects, sutures can be placed individually and tied at the end. Sutures may need to be placed around lower ribs in large defects.

7. If the defect is too large for primary closure, a prosthetic patch of artificial or natural graft material is fashioned in the size and shape of the defect, allowing for a small amount of curvature. The choice of material will depend on local availability, but can include polypropylene, Dacron®, Gore-Tex® (polytetrafluoroethylene), Surgisis®, and Permacol®. The patch is sutured in place with nonabsorbable sutures in a manner similar to that described above; again, the lower ribs may need to be used to anchor the stitches. A chest drain is not mandatory, but is used by some judiciously.

8. If artificial material is not available, a muscular graft (e.g., abdominal wall or a lattissimus dorsi graft) is created to close the defect.

9. Abnormalities of rotation can be associated with CDH, and if a narrow midgut mesentery is present, a Ladd's procedure is performed.

10. Abdominal wall closure can be difficult due to the increased tension caused by return of the intestines into the abdomen. Occasionally, to avoid a tight abdominal wall closure with the consequences of respiratory compromise and abdominal compartment syndrome, the abdomen may need to be closed with a patch.

11. A postoperative chest x-ray is performed to check the position of the diaphragm.

Laparoscopic and thoracoscopic approaches for CDH repair have been described by some centres in select cases. These approaches are suitable for specialised personnel in experienced centres, as they can impose further physiological stresses on the infant.

## Postoperative Complications

Bleeding due to trauma to liver or spleen can occur intraoperatively and should be anticipated with cross-matched blood. Trauma to the intestines, leading to perforation and peritonitis, is also possible.

Postoperative pleural effusion is expected in the immediate postoperative period. Persistence of this can impair lung expansion and weaning off the ventilator. This complication is increased if a hernia sac is not identified and left in situ. The sac then will act as a compartment for fluid to accumulate. Intraoperative excision of the hernia sac, if present, is therefore the best prevention. Management usually consists of inactivity to allow the fluid to resorb. In those cases where this is delayed, thus causing respiratory symptoms or delayed recovery, drainage via a chest drain may be required; this is seldom necessary.

Mediastinal shifts can occur in the postoperative period, as pressure and volume changes due to reducing the abdominal contents ensue. The mediastinal shift induced by CDH does not usually shift back to the central position immediately, but does so slowly. The space is initially filled by air (Figure 45.6) and later on by fluid.

Misinterpretation of the postoperative changes can lead to unnecessary insertion of a chest drain. This can cause large changes in volumes and pressures, with consequential changes in lung expansion, resulting in a true pneumothorax (especially in the contralateral lung, which will then require drainage).

An incisional hernia can occur in patients with tight abdominal wall closure and those with patch closure of the abdominal wall. Semielective or elective repair after a period of stabilisation and growth is advisable.

Recurrence is seen in 5–15% of patients.[9,10] The incidence of recurrence is higher in patients in whom a patch repair is needed (up to 50%) and in those in whom the closure is under tension. The incidence of recurrence is reported to be lower in patch repair using biological-based material (e.g., Permacol);[11,12] however, this is not a universal finding.[13] Management of recurrence is surgical, with principles similar to those for primary surgery. With large defects and those with multiple recurrences, the need for muscle-based (e.g., abdominal wall or latissimus dorsi) flaps[14,15] should be considered.

Gastro-oesophageal reflux is seen in 50–90% of patients,[9] and should be treated as in any other patient. Overall, however, the requirement for surgical fundoplication is higher than in normal children.

Poor feeding and growth are also seen in some (sometimes needing gastrostomy placement).

Intestinal obstruction caused by adhesions can occur,[10] and initially is treated conservatively or operatively, depending on clinical status.

Chest wall deformities can occur in the forms of pectus carinatum (approximately 30%) and scoliosis (20%).[9] The incidence is higher in those patients with a large defect requiring a tight closure, or those

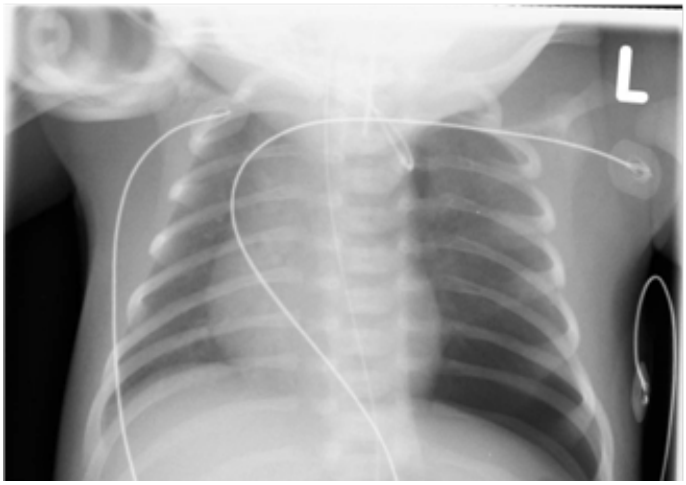


*Figure 45.6: Early postoperative chest x-ray in same patient as in Figure 45.2. The lower right chest cavity is filled with air and a small amount of fluid. There is still some mediastinal shift.*

requiring a patch repair. Management is usually conservative, as these deformities tend to be mild. Operative intervention is reserved for those with severe deformities.

Chronic lung disease is common in severely affected patients.[9] A mild restrictive pattern of lung function is seen in most patients, but is not necessarily associated with clinical symptoms. Alveolar growth continues up to 8 years of age, and children can outgrow any mild restrictions to exercise tolerance and susceptibility to chest infections. An increased incidence of asthma is seen. Those with severe neonatal lung disease develop chronic lung disease. Continued pulmonary hypertension into infancy and childhood are associated with poor outcome. There is a late mortality, due to chronic lung disease and associated or secondary cardiac dysfunction, which can be as late as 4 years of age.[16]

Neurodevelopmental delay and hearing loss are nonsurgical complications that are consequences of poor oxygenation; they are twice as common in children who receive ECMO support.[9,16]

### Prognosis and Outcomes

A summary of the major significant outcome measures and their main determinants (in parentheses) follows.

- death (pulmonary hypertension, associated cardiac anomalies, chromosomal abnormalities, early severe disease);
- neurological impairment and hearing loss (pulmonary hypertension, size of defect, early severe disease);
- chronic lung disease (pulmonary hypertension, size of defect, early severe disease); or
- recurrence (size of defect, need for patch repair).

Thus, the main adverse determinants of outcome seem to be associated chromosomal and cardiac anomalies, severity of the pulmonary hypertension, and size of the defect.

### Prevention

There are no known preventive measures for CDH.

### Ethical Issues

Two main ethical issues surround the management of CDH. Both are outside the scope of this text, but are discussed here briefly. The first is the indication and benefit of any antenatal intervention in the foetus. Antenatal plugging of the trachea is theoretically advantageous by allowing increased foetal lung growth.[17] The indications for intervention are not clear from the research done to date. It is said that infants with adverse features on antenatal scan could benefit from plugging. Intervention is possibly too late to affect the developmental consequences at this stage, however; this may be borne out in the lack of convincing benefit to date.[2] The resources and personnel necessary to run such a programme (or even research into it) are huge, and the debate on cost versus benefit is likely to continue.

Second, the use of ECMO as "rescue therapy" for infants with severe lung disease remains contentious. In one large trial in the United Kingdom,[16] the benefit of ECMO on survival in CDH could not be established. A meta-analysis of randomised trials also failed to show a long-term benefit (late mortality was similar in ECMO and non-ECMO CDH patients).[18] Furthermore, the morbidity induced in those that do survive (related complications included intracranial infarct or bleed, major bleeding, seizures, and infection) is significantly high[16] and can be costly. Around one-fifth have severe neurodevelopmental problems.[16] The use of ECMO continues, and its advantages need to be continually investigated to resolve this issue.

## Eventration of the Diaphragm

Eventration of the diaphragm is defined as an abnormal elevation of an otherwise intact diaphragm due to poor or absent musculature. Although some of the mechanical effects are similar to those of CDH, the incidence of pulmonary hypertension is low and the degree of associated pulmonary hypoplasia is minimal. Thus, the presentation is less dramatic and usually somewhat delayed and the outcome is significantly better.

### Demographics

Like CDH, eventration of the diaphragm is more common on the left. Bilateral lesions are rare. There is no gender or race predilection.

### Aetiology/Pathophysiology

Eventration of the diaphragm is thought to result from failure of myoblastic transformation of the diaphragm or faulty ingrowth of muscle into the dome during embryogenesis. The involved hemidiaphragm is therefore inactive and demonstrates paradoxical movement with respiration.

Although the elevated diaphragm can lead to lung compression and hypoplasia with associated pulmonary vasculature hypertension, as in CDH, this complication is uncommon or mild in eventration. The incidence of other anomalies is low.

Paralysis of the diaphragm due to phrenic nerve palsy or traumatic/iatrogenic phrenic nerve damage can give a picture similar to eventration. The distinction is sometime difficult in those with a potential cause (birth trauma, cardiac/thoracic surgery). To some extent, the distinction is not important because the treatment, *in the symptomatic child,* is usually the same.

### Clinical Presentation

#### History

Respiratory distress, tachypneoa, and cyanosis may be present in the early neonatal period in severe cases of eventration of the diaphragm. Presentation is more often less dramatic and later than for those with CDH. Due to the limited respiratory reserve, poor feeding or sucking, associated with tiring, is common. Failure to thrive may be the presenting complaint due to poor feeding. Vomiting may be the presenting complaint. Failure to recover from a lower respiratory tract infection or recurrent infections may prompt a chest x-ray that brings the diagnosis to light.

#### Physical

The physical findings may be minimal. There may be signs of respiratory distress. Decreased air entry may be present in both lungs but more marked on the affected side. The cardiac impulse may be shifted away from the affected side.

### Investigations

A plain chest x-ray is suggestive of the diagnosis in most cases. Signs on the radiograph are that the right hemidiaphragm is more than two rib spaces higher than the left (Figure 45.7), or the left is more than one rib space higher than the right (Figure 45.8).

It is sometimes difficult to distinguish the radiological picture of eventration of the diaphragm from that of CDH. Unlike the case with CDH, however, there is usually a suggestion of a thin rim of diaphragm.

Fluoroscopy is diagnostic in most cases. Paradoxical movement of the affected diaphragm is seen during screening. (This sign is lost in patients who are ventilated.)

Ultrasound screening to demonstrate paradoxical movements can also be used to make the diagnosis, but this process is less sensitive than fluoroscopy, mainly due to the inability to see both diaphragms simultaneously.

### Management

Patients who are asymptomatic or patients who improve without intervention may be treated conservatively. Conservative treatment for asymptomatic cases suspected to be due to phrenic nerve injury can also be advocated, with hope for recovery if possible. Operative management is the treatment of choice in symptomatic cases.

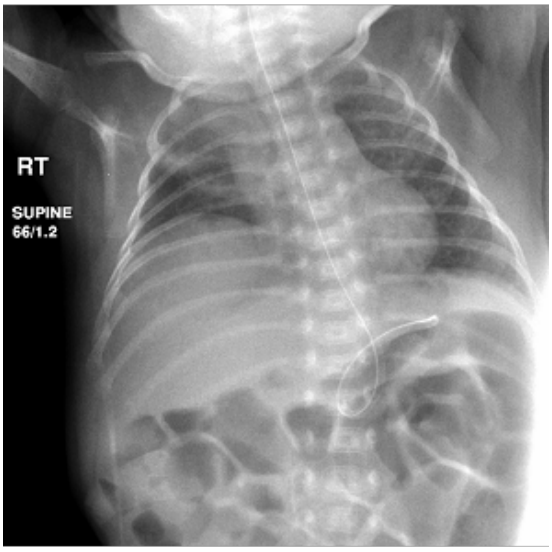


*Figure 45.7: Right-sided eventration of the diaphragm.*

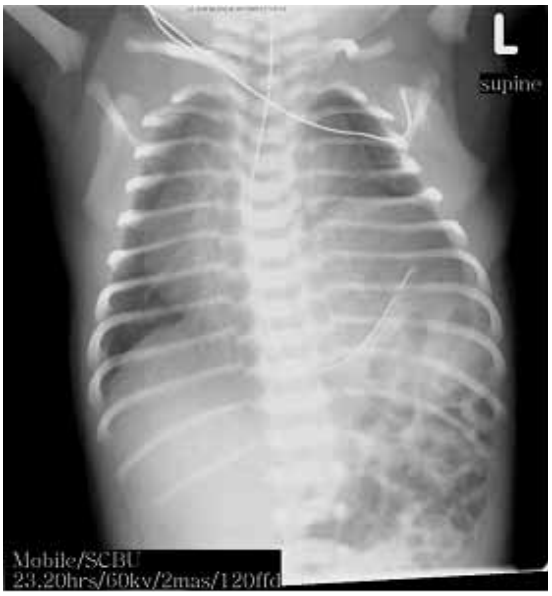


*Figure 45.8: Left-sided eventration of the diaphragm.*

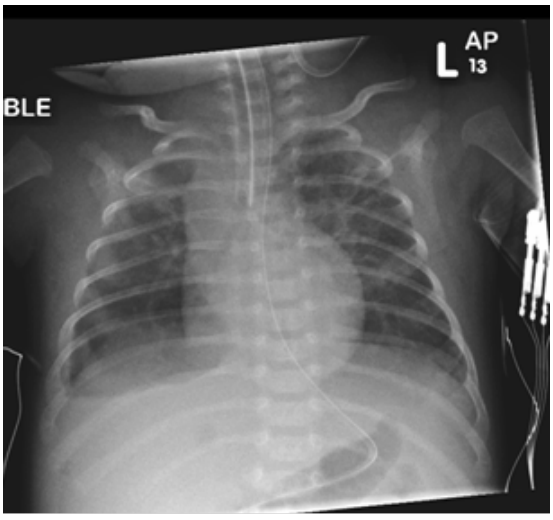


*Figure 45.9: Postoperative chest x-ray after right-sided eventration repair for the same patient as in Figure 45.7.*

## Surgery

For left-sided lesions, the approach can be either abdominal or thoracic. For lesions on the right side, a thoracic approach is preferential. The thoracic approach is through a posterolateral 7th to 9th rib space. The abdominal approach is usually subcostal. Bilateral cases can be approached through a transverse upper abdominal incision. A thoracoscopic approach may be used if skills and resources are available.

The key features of the operation are:

- Confirmation of the diagnosis (versus CDH).
- Plication of the diaphragm by using several rows of pledgeted nonabsorbable sutures to obtain a relatively flat diaphragm.
- Bites of the diaphragm are taken at suitable intervals (~1 cm) in a radial fashion (usually, three or four rows of sutures are needed).
- These sutures can be placed individually without tying, and tied sequentially at the end.
- Identification and avoidance of the branches of the phrenic nerve, if possible.
- A chest drain may be left after a thoracic approach, although this is not mandatory.

A postoperative chest x-ray is suggested to check the position of the diaphragm (Figure 45.9).

## Postoperative Complications

There are relatively few postoperative complications with this surgery.[19] Trauma to the intestines and liver during a thoracic approach is avoided by careful placement of sutures into the diaphragm. Postoperative pneumothorax is also uncommon, and usually resolves with chest drainage. Adhesive intestinal obstruction is possible after an abdominal approach. Recurrence is possible, but is much less common than in CDH.

## Prognosis and Outcomes

The prognosis is very good in the absence of other anomalies. Respiratory mechanics are improved by plication, with increased tidal volume and vital capacity and improvement of symptoms. In a follow-up study 1 to 5 years postplication, there was no recurrence of symptoms,[19] with only one of nine patients having an elevated diaphragm. In the rest, the diaphragm was flat but immobile.

## Prevention

Avoidance of the phrenic nerve during surgery and procedures that put it at risk is preventive in cases due to phrenic nerve injury.

## Evidence-Based Research

Table 45.1 presents a meta-analysis that evaluates the use of ECMO in infants with CDH.

*Table 45.1: Evidence-based research.*

| | |
|---|---|
| **Title** | Extracorporeal membrane oxygenation in infants with congenital diaphragmatic hernia: a systematic review of the evidence |
| **Authors** | Morini F, Goldman A, Pierro A |
| **Institution** | Great Ormond Street Hospital for Children NHS Trust, London, UK |
| **Reference** | Eur J Pediatr Surg |
| **Problem** | The aim of this study was to evaluate the evidence supporting the use of extracorporeal membrane oxygenation (ECMO) in infants with congenital diaphragmatic hernia (CDH). |
| **Intervention** | A meta-analysis of randomised controlled trials (RCTs) comparing ECMO and conventional mechanical ventilation (CMV) |
| **Comparison/ control (quality of evidence)** | Meta-analysis |
| **Outcome/ effect** | The early mortality was significantly lower with ECMO compared to CMV (RR 0.73 [95 % CI 0.55-0.99]; $p < 0.04$); however, late mortality was similar in the two groups (RR 0.83 [0.66-1.05]; $p = 0.12$). |
| **Historical significance/ comments** | Nonrandomised studies suggest a reduction in mortality with ECMO. However, differences in the indications for ECMO and improvements in other treatment modalities may contribute to this reduction. The meta-analysis of RCTs indicates a reduction in early mortality with ECMO but no long-term benefit. |

## Key Summary Points

1. Congenital diaphragmatic hernia is associated with a relatively high mortality related to the associated pulmonary and cardiovascular abnormalities present, and carries some long-term morbidity in most cases. However, self-selected patients who present late have little long-term morbidity.
2. Ventilatory and support mechanisms for patients with congenital diaphragmatic hernia have evolved significantly, but with minimal impact on survival in the severe cases.
3. At present, advanced support for congenital diaphragmatic hernia using ECMO is costly and does not seem to reduce the long-term mortality; it may contribute to more morbidity.
4. The most important factors influencing outcome and long-term morbidity seem to be associated chromosomal and cardiac anomalies, the severity of the pulmonary hypertension, and the size of the defect.
5. Surgery for congenital diaphragmatic hernia is simple in most cases, but can be technically demanding in those with a large defect, requiring knowledge of methods available for secondary closure. Surgery for recurrence can also be demanding, calling for advanced flap procedures.
6. Other surgical procedures for gastro-oesophageal reflux and feeding difficulties or other complications may be required.
7. Unlike congenital diaphragmatic hernia, eventration of the diaphragm is not usually associated with severe morbidity and mortality in most cases. Surgical correction of symptomatic cases is most often rewarded with prompt recovery with little long-term outcome.

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