

NCCU CLINICAL GUIDELINES  
SECTION: 2

RESPIRATORY PROBLEMS AND MANAGEMENT

Section 2: Respiratory problems and management  
Congenital diaphragmatic hernia (CDH)  
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Neonatology Clinical Care Unit Guidelines  
King Edward Memorial/Princess Margaret Hospitals  
Perth Western Australia  
Authorisation & review by  
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## CONGENITAL DIAPHRAGMATIC HERNIA (CDH)

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CDH is a developmental defect of the diaphragm, which may be unilateral or bilateral, partial or complete, which allows the abdominal contents to herniate into the chest.

### EPIDEMIOLOGY

CDH occurs in approximately 1/2500 live births in Western Australia.

### PATHOPHYSIOLOGY

Usually, the defect is in the posterolateral (Bochdaleck) segment of the diaphragm, usually on the left (80%). This is due either to failure of closure of the pleuro-peritoneal canals, or failure of migration of the post-hepatic mesenchymal plate. Both lungs are hypoplastic, especially the ipsilateral lung.

The pulmonary parenchyma is poorly developed with abnormal structure and decreased numbers of alveoli. In addition the pulmonary vasculature is hypoplastic and the medial muscle layers of the pulmonary arterioles are unusually thickened, making them particularly susceptible to vasospasm during periods of hypoxia, hypercarbia and acidosis. These structural and physiological abnormalities result in highly varying amounts of pulmonary hypertension, some of which is responsive to treatment and some of which is not! The result is marked shunting of blood right to left at several levels in the heart.

CDH is frequently associated with other anomalies. 30-50% of fetuses with CDH have another anomaly (30% with a cardiac anomaly) but intra-uterine deaths, terminations and stillbirths reduce this to about 20-30% of live born infants.

**Chromosome, FISH** (e.g. 8p23.1, one of the recurrent chromosomal causes of CDH) **and micro array studies** should be considered in all babies with CDH, especially in any infant with dysmorphisms and or associated anomalies. There are a number of non-chromosomal causes to consider e.g. CHARGE, overgrowth conditions (e.g. Beckwith or Simpson-Golabi- Behmel), Cornelia de Lange syndrome, Donnai-Barrow etc Every infant with a CDH should have a head, heart and renal ultrasound performed. A genetics referral will depend on the presence or absence of associate anomalies.

### CLINICAL PRESENTATION

#### ANTENATAL

An antenatal diagnosis is made in >75% of all cases of CDH. Bowel loops and/or liver are seen within the chest with a shift in the heart. Polyhydramnios is frequently present and in some cases hydrops develops. All infants with CDH should be delivered in a tertiary centre with appropriate obstetric and neonatal care.

## POSTNATAL

The classic presenting picture of the more common left sided CDH is of a newborn with severe respiratory distress and a scaphoid abdomen with breath sounds reduced on the left and the heart sounds best heard on the right side of the chest.

About 5% of CDH will present at >24 hours of age with mild tachypnoea, or later still with failure to thrive, recurrent chest infections, pleural effusions or as an incidental finding on CXR.

## OUTCOME

Survival depends primarily on the degree of pulmonary hypoplasia and the amount of fixed pulmonary hypertension and secondly on the severity of associated anomalies. Many centres around the world are now reporting 80-90% survival in isolated CDH. However, the survival rate varies according to the CDH population being treated (see Colvin et al). Increasing survival has also seen increasing morbidity i.e. growth and neuro-developmental delay; 'chronic lung disease'; long term feeding difficulties; and often recurrent herniation.

The past survival of children born with CDH in WA was <50%. This survival rate has steadily increased. Over the last 12 years, 1/1/02 to 31/12/14, there were 82 babies born with CDH in WA: 62 at KEMH and 20 born outside KEMH and transferred to PMH. Overall 18 babies died, giving a 78% survival rate. Of the 70 babies arriving at PMH, from all sources, 63 (90%) survived. Overall, between KEMH and PMH, 53/63 (84%) left and only 11/19 (58%) babies with right CDHs survived.

## INVESTIGATIONS

CXR is diagnostic in most infants showing air-filled loops of bowel in the hemi thorax. Occasionally CDH may appear as a solid mass on the first chest X-ray after birth; the loops of bowel may initially be fluid filled on the left or the liver may be the only herniated organ on the right. The differential diagnosis includes CCAM (see next page).

## MANAGEMENT

While many ante natal factors have been assessed to try and predict post natal survival, e.g. degree of polyhydramnios, the size of the pulmonary arteries etc none have been consistently accurate.

Timing of delivery is determined by obstetric factors other than the CDH. There is no evidence that the type of delivery influences the outcome of CDH.

## LABOUR WARD MANAGEMENT

- In known or suspected cases the immediate priority is to intubate the trachea. Bag and mask ventilation should be avoided to minimise gas distension of the intra thoracic bowel.
- If the heart rate is good and colour/SaO<sub>2</sub> is improving, keep inspiratory pressures low and ventilate at a rate of 30-60bpm and watch chest wall movement. Try not to over ventilate.
- Monitor pre-ductal SaO<sub>2</sub>: aim for > 85 %.
- Insert a large bore Nasogastric tube (8 or 10 FG) and keep on open drainage.

## NICU MANAGEMENT

A major reason for the improved outcome of babies with CDH has been the adoption of a **Lung Protective Strategy**. Historically CDH infants were treated with high-pressure ventilation and high rates to maintain normoxaemia and normocarbica, often hypocarbica to relieve pulmonary hypertension. This practice frequently resulted in severe barotrauma and poor outcome.

The aim of the **Lung Protective Strategy** is to minimise barotrauma by accepting mild hypoxemia ( $\text{SaO}_2 > 85\%$ ), hypercarbia ( $\text{PaCO}_2$  45-55) and acidosis ( $\text{pH} > 7.28$ ).

**Monitor pre-ductal  $\text{SaO}_2$**  as this reflects cerebral oxygenation: aim for  $> 85\%$ . Simultaneous Pre and **Post ductal  $\text{SaO}_2$**  measurements can be a useful guide to the degree of ductal right to-left shunting.

A **large-bore nasogastric tube** (8 or 10 FG) should be passed to decompress the stomach and small bowel. A **Replogle tube** on continuous suction can be useful if the thoracic bowel cannot be deflated.

All infants should have an **arterial line**, access for multiple IV infusions and, a double lumen **central venous line** with CVP measurements (5-8mmHg) in ideal circumstances

All infants should be well **sedated** with [Morphine](#) plus or minus [Midazolam](#). Consider giving a bolus dose of morphine and or Midazolam a few minutes before any disruptive procedures, such as suctioning etc. [Fentanyl](#), 2-3 microgram/kg IV immediately prior to a disruptive procedure is also an option in very fragile babies.

**Note:** Excessive sedation can result in systemic hypotension.

A short period of partial or complete **paralysis** with [Vecuronium](#) or [Pancuronium](#) can be helpful in those babies who are struggling and have severe lung disease.

There is no clear evidence favouring either **HFOV or Conventional Ventilation (CV)** in the management of CDH. There is some evidence from historical reviews favouring the use of HFOV, but there are many confounding issues with these studies. In general, however, HFOV is preferred to CV in infants with significant respiratory failure. The optimal starting MAP on HFOV will vary between infants: start at, or 1-2 cm  $\text{H}_2\text{O}$  above CV MAP and then slowly increase the MAP until an acceptable  $\text{SaO}_2$  is achieved. Further increases in MAP, as per Lung Recruitment Strategies, should be used very carefully in babies with CDH. Remember you are ventilating the infant on 'one lung'. Vary the amplitude (shake) according to the  $\text{PaCO}_2$ . Use frequencies of 8-12 Hz with HFOV. Consider switching to HFOV if requiring PIP  $> 25$  cm $\text{H}_2\text{O}$  on CV and try to keep MAP to  $< 16$  cm $\text{H}_2\text{O}$ .

In CV, SIPPV (back up rate of 45-60bpm) with VG (it is reasonable to start with 4ml/kg and vary with chest wall movement and ABGs) seems an appropriate strategy in a well-sedated baby. Care should be taken as the baby improves pre operatively not to allow the baby to breath at 'CPAP pressures' for too long when extubation is not expected, as this will most likely increase the baby's work of breathing and cause the baby's oxygen requirement to 'flip flop'.

With either CV or HFOV, as conditions improve it is very important to make small changes to ventilation, especially with MAP, again in order to avoid 'flip-flop'.

Whilst there is no evidence that **iNitric Oxide (iNO)** improves outcome in CDH, all infants with CDH and respiratory failure should be started on iNO. Initial dose 10-20ppm. While there may not be an obvious improvement with iNO, clinical practice suggests that iNO smooths out the highs and lows of pulmonary hypertension. Subsequently an infant can show exquisite sensitivity to even minor downward changes in iNO, suggesting that iNO has been effective in the management of pulmonary hypertension.

Other pulmonary vasodilators have been used to treat pulmonary hypertension. These include: **Epoprostenol (PG I2)** given either by IV infusion (easily available now) or by inhalation; **PG E1**;

**Adenosine infusion; magnesium infusion; and NaHCO<sub>3</sub> infusion.** Several reports have shown that **oral Sildenafil** can be used during the acute management of pulmonary hypertension, in babies not being fed, with apparent good effect. **Note:** Both PG E1 and PG I2 can cause significant systemic vasodilatation and therefore hypotension.

Routine **surfactant** use in CHD has been controversial with limited studies on small numbers of babies showing no advantage to routine use. One study even suggesting a disadvantage. Most clinicians would use surfactant in preterm infants with CDH. The logical choice of surfactant would be [Curosurf](#), given the smaller dose volume.

Some degree of **inotropic and or vasopressor support** is frequently required. This may vary from [Dobutamine](#) in babies with normal BP but suboptimal cardiac output; to the use of [Dopamine](#) (or adrenaline or noradrenaline) in hypotensive babies. A combination of these agents is frequently required. [Milrinone](#) can be a useful afterload reducing agent. The use of these agents should be guided by a combination of clinical, blood gas and echocardiographic findings. Optimal Mean Blood Pressure (MBP) in CDH can be difficult to determine; a MBP of 45-55mmHg in a near term infant would be optimal, using blood gas, lactate, urine output (min. of 0.5ml/kg/hr) and peripheral perfusion as a guide. Some clinicians will aim for higher MBPs in order to exceed the supra systemic BPs associated with pulmonary hypertension.

**Volume expansion** is also frequently required, either because of 'third space' losses into the abdomen or systemic vasodilatation or capillary leak syndrome. However, fluid retention and marked peripheral oedema can easily develop and the careful use of **Fruzemide** can significantly improve lung function.

A recent study showed that 67% of babies with CDH are **adrenal insufficient** which resulted in a poorer outcome. Urgent cortisol levels should be checked in every baby with CDH who requires significant cardio-respiratory support. At present a decision to supplement with hydrocortisone can only be made on a case by case basis when all factors are taken into account.

Maintaining **ductus arteriosus patency with PG E1 ([Alprostadil](#))** has been reported as beneficial in preserving right ventricular function. PG E1 is routinely used in all CDH babies at the Royal Children's Hospital, Melbourne (personal communication) until pulmonary hypertension has resolved. A low dose (5-10nanogm/kg/min) is usually sufficient if started early. PG E1 may also assist in directly reducing pulmonary vascular resistance. Systemic hypotension can be a problematic side effect.

**IV [sodium bicarbonate](#)** can be used **to correct metabolic acidosis** after all other factors contributing to the metabolic acidosis are corrected. Generally a mild metabolic acidosis can be accepted, as per the Lung Protective Strategy. However in difficult to control pulmonary hypertension, IV sodium bicarbonate can also be used to achieve a metabolic alkalosis as an aid **to vasodilate the pulmonary circulation:** aim for a pH of approximately 7.45-7.5. Sodium overload can be a problem.

All babies with CDH should have an early **echocardiogram** to rule out congenital heart disease and to assess cardiac function and ductal patency. Repeat echocardiograms may be helpful in monitoring right ventricular function, pulmonary hypertension and ductal patency.

There are many general measures, which are important in the care of infants with CDH.

**Maintenance fluids** should be kept around 60 ml/kg/day in order to reduce water retention and oedema. **Antibiotics** should be given in all cases initially until cultures are clear. **Calcium and**

**magnesium** levels should be monitored and kept within the normal range to ensure optimal cardiovascular function. **Parenteral nutrition** needs to be optimised to prevent catabolism.

## ECMO

A Cochrane review of ECMO in severe respiratory failure (oxygenation index >40) in near term babies, which included CDH, significantly favoured the use of ECMO. However, while the short term outcome of CDH babies was improved, the longer term outcomes were not improved. They concluded that the use of ECMO in severe CDH is unclear (Nov. 2007). Future studies may better define the characteristics of a sub group of CDH babies who would benefit from ECMO.

ECMO is available to neonates on a case-by-case decision through the PMH PICU. Babies who could be considered are those over 2.5-3kg, no significant associated anomalies, who have been stable but then suddenly deteriorate especially in the post op period.

## TRANSPORT OF A BABY WITH KNOWN OR SUSPECTED CDH

Please refer to the [NETS WA](#) Guidelines.

Newborns delivered in peripheral hospitals with suspected diaphragmatic hernia will require immediate transfer to PMH. Those antenatally diagnosed (about 75% of CDH cases in the last 13 years in WA) and born at KEMH will also require transfer to PMH at some stage. The timing of this transfer has been the subject of much debate. The goal should be to transfer all babies to PMH for surgery when they are stable. While not wanting to miss a 'window of opportunity' to transfer such babies, there is no indication to transfer a newborn with CDH urgently to PMH.

A **stable** baby on high frequency ventilation and nitric oxide may be considered for transfer to PMH. Whilst HFOV cannot be administered on transport, nitric oxide can. A trial of conventional ventilation is warranted, & if tolerated without an unacceptable rise in ventilatory requirements, transfer can be arranged.

## SURGERY FOR CDH

The **timing of surgery** has been the subject of much debate but very little formal study. A Cochrane review comparing early (<24hrs) and late surgery could not find any advantage with either approach. Only 2 studies were included in this review, both with small numbers. Our current approach is to await an almost complete return to normal cardiorespiratory requirements. However this may be too strict a requirement. Infants do not need to be off iNO or HFOV for surgery. Recently a 1.5 kg 30 week gestation baby with a large defect, who was stable on approx 10ppm of iNO and on HFOV had a repair, with a patch, performed on the ward with an excellent result and subsequent rapid weaning of his support. For a variety of good reasons CDH surgery is not available at KEMH.

## POST OPERATIVE CARE

All babies should have a CXR (AP and Lat) immediately post op. There will always be a large ipsilateral "pneumothorax" secondary to the thoracotomy. This is rarely drained. This space quickly fills with fluid and expanding lung over the first post op days. The effusion does not usually require drainage but if the child is deteriorating and the effusion seems to be preventing the lung from expanding, then a trial removal of a small amount of the effusion should be considered; the fluid

should be sent to micro and biochem exam to exclude chyle and infection. A pneumothorax or effusion on the contra lateral side to the hernia is never normal and should be treated accordingly.

General management should proceed as per pre op management. Mean airway pressure should be weaned first, followed by iNO. SIPPV or SIMV with VG work well; do not allow a baby to stay on “CPAP pressures” on VG for a long time without a plan to extubate as this will tire the baby unnecessarily. PSV can be a useful weaning strategy as pressure support is gradually decreased. Fluid overload and gross oedema is common in the post op period; the baby’s respiratory requirements can be significantly improved with the judicious use of diuretics.

The use of nasal CPAP (5-8 cm H<sub>2</sub>O) has significantly reduced the need for long-term ventilation in many babies with CDH. The increasing survival of ‘borderline babies’ has seen more babies needing long term CPAP and oxygen.

Tolerating enteral feeding can be problematic. Gastro-oesophageal reflux is frequently present and often treated with thickened feeds and proton pump inhibitors (both of uncertain value and in the case of thickening agents potentially harmful). Continuous feeds (24-30 calorie) can be a useful strategy to ensure adequate caloric intake. Infant positioning, prone with slight head up, can also be helpful.

Recurrent herniation can occur, especially in babies who have a patch in situ. This can sometimes be difficult to diagnose; ultrasound can be useful.

Unexpected respiratory deteriorations can be due to many reasons including: sepsis; aspiration, effusions; pneumothorax; atelectasis with tiredness needing CPAP; and re herniation.

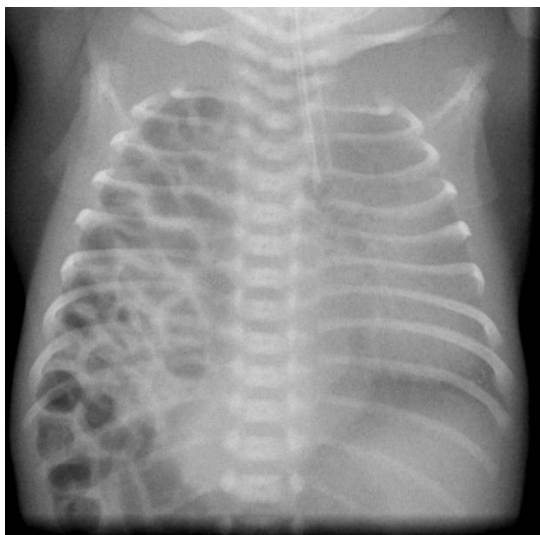
**Discharge planning** is often simple with only neonatal and surgical outpatients required but it can be complex with multiple sub specialists involved plus the need for a general paediatrician for longer term follow up and coordination of care. All babies should be enrolled onto Developmental Follow up. All babies should be assessed by a Respiratory Physician before discharge.



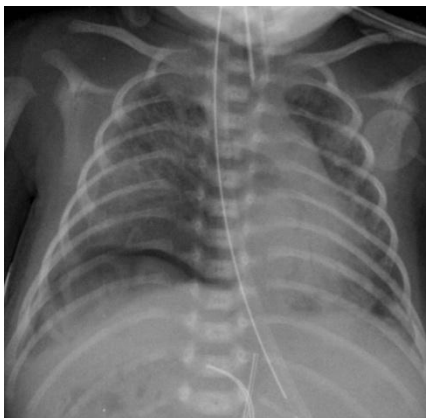
Left CDH. ETT and UAC are in good position. Difficult to say where the UVC ends ?Liver. Note no gastric tube resulting in significant aeration of intra thoracic bowel loops.



Lateral of above patient.



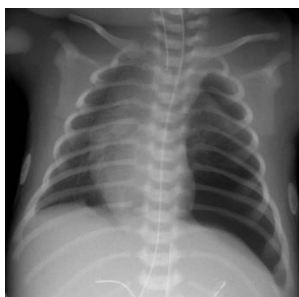
Right CDH.



Post op CXR of above Right CDH patient. Note obvious pneumothorax.



Post op Right CDH as above. Pneumothorax filling up with fluid.



Immediate post op Left CDH. Looks like an obvious Left tension pneumothorax. Does not require an ICC.

## REFERENCE

Trends in treatment and in-hospital mortality for neonates with congenital diaphragmatic hernia  
Hagadorn, et al. Journal of Perinatology (2015), 1–7