Title:	Guideline for the Management of Babies with Congenital Diaphragmatic Hernia (CDH)
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Management of Babies with Congenital Diaphragmatic Hernia (CDH)

This document has been developed by consensus across various neonatal and paediatric specialist teams across the North West Region and is based primarily on two international consensus statements [1, 2].

Antenatal management

Due to improved second trimester ultrasound, CDH has become a prenatal diagnosis in the main. On antenatal detection of CDH, detailed expert assessment will be performed to determine the outcome of the defect, the observed/expected lung-to-head ratio and the position of the liver, in addition to ruling out other associated congenital anomalies or chromosomal anomalies. Associated chromosomal/genitourinary anomalies are found in 25% cases, and cardiac anomalies in 20% cases.

When a woman is identified as potentially having a baby with a CDH, they should ideally have an initial assessment by the fetal medicine team as close to 20 weeks as possible. The following information should be gathered:

1. Description of the defect:
    a. Location
    b. Observed/expected lung-to-head ratio (LHR)
    c. Liver position (intra-thoracic or intra-abdominal)

2. Exclude associated anomalies
    a. Cardiac
    b. Genitourinary
    c. Syndromes/chromosomal anomalies
    d. Amniocentesis for CGH-Array

3. Discussion of options
    a. Prognosis based on the nature of the defect
    b. Care during pregnancy & post-delivery
    c. Option to continue care or offer termination

The following features detected antenatally are associated with a poor prognosis and should guide antenatal counselling [3,4]:

- Right sided hernia
- Intrathoracic position of liver
- Associated congenital heart disease
- Chromosomal abnormality
- Low observed/expected lung volume ratio on antenatal US/fetal MRI

Following a diagnosis of a CDH, the specialist fetal medicine centre will organise a joint

counselling session for parents with a fetal medicine consultant, paediatric surgeon, neonatologist and any other relevant allied professionals such as the Palliative Care team. This first joint counselling session at LWH Fetal Centre or SMH FANS clinic will typically be around 28 weeks' gestation and include a repeat scan to assess lung:head ratio, overall growth/Doppler studies and a fetal echocardiogram. The outcome of the discussion must be documented and copies sent to the parents.

Antenatal care should be transferred to one of two tertiary centres (SMH or LWH). Delivery of these babies should also take place at the same tertiary surgical centre. Babies delivered at LWH will be transferred to Alder Hey Children's Hospital for surgery. An experienced tertiary centre is the optimal environment for the delivery and neonatal treatment of prenatally diagnosed CDH [5,6]. If unexpected delivery takes place outside of these units, Connect NW should be notified at an early stage in order for preparations to be made for immediate dispatch once the baby is born.

Antenatal steroids should be given in cases of anticipated delivery at < 34 weeks' gestation [1,2].

Management at delivery and stabilisation prior to surgery

Delivery Room

An experienced neonatal team should be present at delivery including a Consultant, Junior Doctor/ANNP, and two nurses with one experienced in the delivery of babies with CDH. Where possible, the neonatal team should have time to prepare necessary equipment. Basic resuscitation manoeuvres (such as good thermal management) should be followed.

1. Delayed cord clamping should be considered (in accordance with local practice).
2. Large bore NG tube to be passed and kept on free drainage to decompress stomach.
3. Intubate as early as possible to prevent bowel distension and further lung compression.
4. Pre-medication should be considered [7]:
a. LWH: intranasal midazolam/fentanyl for sedation/analgesia
b. SMH: IM atracurium for paralysis
5. Confirm tracheal tube in place using capnography
6. Gentle ventilation should be started, guided by oxygen saturations and heart rate. Aim for low peak pressures [1,2], preferably < 25 cmH20. Initial oxygen to be set at 21% and adjusted according to heart rate and pre-ductal saturations.
7. Site SpO2 probe (ideally on right arm) and aim for pre-ductal saturations of 80-95%.
8. Surfactant should not be routinely administered to term babies with CDH but should be considered in preterm infants [2].

Intensive Care Unit

Respiratory:

Ventilatory management of babies with CDH is a careful balance between ensuring adequate oxygenation and CO2 elimination while minimising ventilator-induced lung injury and oxygen toxicity. This is done through permissive hypercapnia and 'gentle ventilation.' [8].

1. Pre-ductal saturations should guide oxygen therapy in newborns with CDH with alarm limits set at 85-95% for those needing supplemental oxygen
   Supplemental oxygen is often necessary in babies with CDH; while excessive hypoxia can aggravate pulmonary hypertension, oxygen therapy can also be damaging. SpO2 limits can be lowered to 80% with consultant agreement if perfusion is good but high concentrations of oxygen are needed.
   Post-ductal saturations >70% are acceptable but this must be reviewed regularly according to clinical state.

2. Aim to achieve adequate ventilation whilst minimising volume-trauma to the lungs
   Conventional mechanical ventilation is the optimal initial ventilatory strategy. High frequency oscillatory ventilation can be used as rescue therapy if conventional mechanical ventilation fails.

   Pressure-controlled ventilation:
   PIP of 20 cmH20 and PEEP of 3-5 cmH20 are reasonable initial settings. Adjustments should be made based on oxygenation, CO2 clearance and assessment of lung inflation on chest x-ray.

   Volume-guided ventilation:
   TV 4-5 ml/kg, PEEP 3-4 cmH2O with a respiratory rate of 40-50 breaths per minute are reasonable initial settings. Adjustments should be made based on oxygenation, CO2 clearance and assessment of lung inflation on chest x-ray.

   High frequency Oscillatory Ventilation (HFOV):
   HFOV should only be used as recue therapy when conventional ventilation has failed. A change to HFOV should be considered if a baby with CDH requires a PIP >28 cm H2O to clear CO2 or achieve adequate oxygenation.

Cardiovascular:

1. Establish central vascular access

All babies with CDH require central venous and arterial access and umbilical catheters are the recommended method. The typical landmarks for a UVC/PICC may be displaced in babies with CDH making it difficult to identify its true course and location on X-ray. Echocardiography can be used to help identify the line tip's location.

A UAC samples post-ductal blood and in certain situations (e.g. where there is a large pre and post-ductal oxygen saturation difference) a peripheral radial arterial line into the right radial artery (pre-ductal) might be preferable.

2. Maintain adequate perfusion

Perfusion can be affected by cardiac dysfunction secondary to functional hypoplasia of the left ventricle, hypoxaemia and acidosis. Adequacy of perfusion is determined by heart rate, urine output, and lactate levels.

If there are signs of poor tissue perfusion, consider administering up to two boluses of 10 ml/kg normal saline (max. total 20 ml/kg).

3. Maintain blood pressure within normal limits

BP should not routinely be maintained at supra-physiological levels. Aim to maintain BP within the normal range for gestational and postnatal age.

4. All babies should have an echocardiographic assessment within 12-24 hours of birth to:

a. Exclude any structural cardiac anomalies
b. Make a baseline assessment of pulmonary hypertension

Regular functional echocardiography (including assessment of right and left ventricular function) might be useful in management of babies with CDH and should be considered if resources are available.

5. Use of inotropes

Refer to local centre guidance at LWH/SMH.

6. Management of pulmonary hypertension

Refer to local centre guidance at LWH/SMH.

7. ECMO (please also refer to NWNODN ECMO pathway) [1,2]

Early referral and discussion of potential ECMO patients with the ECMO Centre is encouraged for the following reasons:

1. To establish candidacy for ECMO.
2. To enable further optimisation at the referring centre and potentially avoid ECMO.
3. To ensure adequate stabilisation prior to transfer for ECMO.

The following situations should prompt an early discussion with the ECMO Centre:

- Inability to maintain the pre-ductal SpO2 > 85% or post-ductal SpO2 > 70%
- Ongoing need for high pressure ventilation (PIP > 25 cmH20 on conventional ventilation or MAP > 17 cmH2O on HFOV)
- Hypoxaemic respiratory failure with OI > 40 despite optimising conventional ventilation and trial of inhaled nitric oxide[1].
- Hypercapnic respiratory failure with PaCO2 > 8 kPa/pH < 7.15 despite optimal ventilation.
- Inadequate tissue oxygen delivery (indicated by pH < 7.15 or lactate > 5 mmol/L or urine output < 0.5 ml/kg/hr over 12-24 hours)
- Refractory systemic hypotension, resistant to fluid and inotropic therapy.
- Recurrent pneumothorax/air leak.

Exclusion Criteria for ECMO

- Significant coagulopathy or uncontrollable bleeding
- Major intracranial haemorrhage (e.g. > grade 2 IVH) or evidence of brain injury [
- Irreversible lung injury (absence of a period of adequate oxygenation and CO2 clearance despite optimal respiratory management)
- Major congenital/chromosomal anomalies (including cardiac malformation) [1,2]
- Cardiac arrest other than immediately at birth.

Fluids & Nutrition:

- Initial total fluid volumes to start at 60 ml/kg/d and adjusted according to overall assessment of fluid status
- Preoperatively, patients should only receive parenteral nutrition.
- Parenteral nutrition should be started at the earliest opportunity and ideally within 24 hours of birth

Surgery:

Surgical repair of the diaphragmatic defect should ideally be performed after clinical stabilisation [1], defined as follows:

- Maintaining pre-ductal saturation > 85% in ≤ 50% FiO_2 on conventional ventilation with a PIP (on VG-PS/SIMV) of ≤ 28 cm H20
- Ideally off iNO
- Mean arterial BP normal for gestational age
- Serum lactate < 3 mmol/l
- Urine output > 1 ml/kg/hr

The on-call paediatric surgeon should be informed of the baby's birth following admission to the neonatal ICU. Early and regular reviews with the surgical team should take place to ensure pre-operative assessments are performed and to ensure timely transfer/surgery. Information from antenatal MDT assessments and counselling should be available to neonatal, surgical and transport teams.

[1] *Or OI>30 if on HFOV plus inhaled nitric oxide.

A pre-operative MDT involving surgical, anaesthetic and neonatology/PICU teams is an important part of optimal management, especially in babies with cardiorespiratory instability.

Transport:

1. For babies born at LWH, surgery will take place at AHCH and therefore will require transfer by Connect NW (0300 330 9299) to PICU at Alder Hey. A conference call should be arranged between the neonatal, surgical and PICU teams. After surgery, most babies will remain on PICU, rather than being transferred back to LWH, with neonatal input and discharge to neonatal surgical unit when appropriate.

2. If a woman presents at the local hospital with a known fetal diagnosis of CDH, in utero transfer should be arranged where possible by contacting Connect NW. A conference call will be arranged between the neonatal and obstetric staff at the referring and receiving staff. If in utero transfer is not possible then the local team should prepare for the delivery and stabilisation (as set out in this guideline) and contact Connect NW at an early stage once delivery has taken place. A conference call will also be required for any postnatal transfers, where management will be discussed by the local team, Connect NW, surgical and PICU teams. The Connect NW team will be mobilised at the earliest opportunity.

3. The management in a local neonatal unit will normally follow the principles outlined in this guideline with the exception of using HFOV. CNW have the facility to provide HFOV and/or nitric oxide if required during transfer. Most babies will require both sedation and muscle relaxation for the duration of the transfer and ideally should have a minimum of two points of venous access or a double lumen central venous catheter.

Post-operative care:

Care should be provided jointly by surgical and neonatal/paediatric ICU teams. It is important to ensure:

- Ventilation:
  - ventilate on low pressures as per pre-op management
  - non-invasive respiratory support might be needed post-extubation
- Cardiovascular:
  - Aim to maintain blood pressure appropriate for gestational/postnatal age
- Fluids & Nutrition:
  - TPN to be continued postoperatively
  - Anti-reflux treatment when introducing enteral feeds.
- Sepsis:
  - Most babies complete a course of antibiotics post-operatively.

Other specialties

Some of these babies will need input from specialist children's services such as cardiology, dietetics, SALT, and physiotherapy. It is worthwhile considering involving palliative care teams at an early stage, not just for those babies with a poor prognosis, but also for those whose families may benefit from the support that this team can provide.

Follow-up:

All patients will require follow up with the paediatric surgical team and the patient's local neonatologist. Consideration should also be given to respiratory follow up at RMCH or AHCH.

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