

POSTNATAL MANAGEMENT OF CONGENITAL DIAPHRAGMATIC HERNIA GUIDELINE

Background and Antenatal Counselling

- Congenital diaphragmatic hernia (CDH) is rare (1 in 3000 live births), approx. 70% diagnosed antenatally in routine screening, approx. 25% have associated anomalies, overall mortality 30-50% (mostly in the first year of life).
- Disordered lung development from herniation of abdominal organs into the chest results in pulmonary hypoplasia, characterised by abnormal airways and vasculature leading to respiratory insufficiency, pulmonary hypertension and cardiovascular dysfunction postnatally.
- Disease severity decreases with left-sided defect, observed/expected lung-to-head ratio (O/E LHR) >50% and intra-abdominal liver.
- Significant long term morbidity in survivors including impaired respiratory, gastrointestinal, cardiac, and neurological systems; neurodevelopmental delay, and hearing impairment.

Preparation prior to Delivery

- Delivery should be timed to occur in normal working hours. If there is no surgical capacity, then antenatal transfer to another surgical centre should occur
- Complete a STEPP card for high-risk delivery
- Prepare an intensive care cot space with a ventilator and nitric oxide
- Inform the paediatric surgery team of the impending delivery
- Consider discussion with PICU at this stage if the baby is considered to be high risk and possibly requiring ECMO (to facilitate their resource allocation)

List of extra items for delivery room:

Estimate weight using 50th centile for gestation

2x doses of sedation

2x doses of muscle relaxant

2x 5ml 0.9% NaCl flushes

If no IV access is possible, consider IM muscle relaxants if necessary

Delivery Room Management

- Follow NLS Guidance with particular attention to:

Monitoring

- Put saturation probe on the right hand
- Aim for preductal SpO₂ in the delivery room of >80% (>70% and improving) up to 2 h of life

Oxygen

- Start in room air or 30% oxygen based on gestation as per NLS guideline

Sedation and Analgesia/Paralysis

- Do not routinely use muscle paralysis during initial resuscitation - infants with CDH have reduced lung compliance and respiratory function at birth, this is reduced further by the administration of a neuromuscular blocking agent

Intubation

- Avoid using bag and mask or T-piece ventilation as it may cause stomach distension and limit lung expansion

- In infants predicted to have mild pulmonary hypoplasia (e.g. left-sided defect, O/E LHR >50%, intra-abdominal liver), a trial of spontaneous breathing could be considered to reduce ventilator-induced lung injury
- In higher risk infants with prenatally diagnosed CDH, intubate immediately after birth
- The carina is displaced upwards in infants with CDH, consider using a shorter ETT length, use a length of 5.5cm + weight (oral ETT) to prevent right main bronchus intubation with subsequent pneumothorax
- Premedication should be given before intubation where possible as it improves intubation success rates, however do not mask ventilate whilst obtaining IV access, try to intubate without drugs
- If intubated, keep peak inspiratory pressure as low as possible, ideally <25 cm H₂O

Decompress the Stomach

- Prevent bowel distension which may increase lung compression; immediately after birth place a large bore (8 or 10 Fr) orogastric or nasogastric tube with intermittent or continuous suctioning

Surfactant

- Do not routinely use surfactant in term or late preterm newborns as endogenous surfactant amounts are likely to be appropriate to lung size in CDH infants

Management in the NICU

Ventilation Management

- A chest x-ray should be obtained as soon as possible to assess the newborn's initial condition
- A lung protective ventilation strategy should be used. In general use:
 - Volume targeted ventilation, limitation of PIP to <25 cmH₂O, PEEP 3-5 cmH₂O, ventilator rate 40-60/min
- Aim to minimize lung damage so accept:
 - Preductal SpO₂ of 85-95%, post-ductal saturation above 70% and PaCO₂ 6.9-9.3 kPa if pH allows
 - In the first 2 hours of life, pre-ductal saturations as low as 70% are acceptable if they are improving without ventilation changes, if pH >7.2, lactate <5mmol/L, urinary output >1ml/kg/h and if PaCO₂ <8.6 kPa
 - Permissive hypercapnia and the avoidance of high airway pressures have been reported to decrease mortality in infants with CDH
 - If a PIP of >28 cmH₂O is necessary to achieve the above PaCO₂ or saturation levels, consider high-frequency oscillatory ventilation (HFOV) or ECMO.

Vascular Access

- An arterial line should preferably be inserted into the right radial artery in order to measure preductal PaO₂ (which reflects cerebral oxygen delivery). Alternatively an umbilical arterial catheter (UAC) may be sited, however this will reflect postductal PaO₂.
- Repeated vascular access attempts may cause agitation and worsening pulmonary vasoconstriction.

Fluids and Feeding

- Pre-operatively, infants should have no enteral feeds and should only receive parenteral nutrition, if they are likely to be continuously without enteral feeds for more than 5 days.

Sedation and Analgesia

- Sedation should be provided to all mechanically ventilated infants with CDH until weaning from ventilation as agitation may lead to increased pulmonary vasoconstriction.
- Neuromuscular blockade is associated with hypoxaemia and impaired respiratory function. Deep sedation and neuromuscular blockade should be carefully considered only in neonates with high ventilation or oxygen requirements.
- Blood pressure should be closely monitored for potential haemodynamic side effects.

Haemodynamic Support

Blood Pressure Control/Perfusion Support

- Do not increase blood pressure to supra-normal levels if pre-ductal saturation remains above 80%.
- See otherwise ‘Management of Circulatory Failure’ and ‘Management of Persistent Pulmonary Hypertension (PPHN)’ Guideline for further guidance.
- Consider optimising Haemoglobin, Calcium and Magnesium.
- There is increased risk of pulmonary oedema with fluid resuscitation, as the left ventricle may be smaller and less compliant.
- There is an association between CDH and catecholamine-unresponsive systemic hypotension and adrenal insufficiency - hydrocortisone can be considered in the early phase of hypotension if other treatments have failed.
- An echocardiogram should be performed if poor perfusion continues, blood pressure remains below normal values for gestational age, or pre-ductal saturation is <80%.

Echocardiography

- Perform an echocardiogram as soon as possible to ascertain cardiac structure, assess ventricular function, measure pulmonary artery size, assess for ductal and intracardiac shunting and determine the amount of pulmonary hypertension.
- Perform a follow-up echocardiogram at 2-3 weeks of life to check for pulmonary hypertension as there is an association between CDH and persistent pulmonary hypertension beyond 14 days of life and death.

Management of Pulmonary Hypertension

- Use iNO:
 - to treat confirmed supra-systemic pulmonary hypertension without left ventricular dysfunction where lung recruitment is adequate
- OR
- If preductal saturation is <85% and/or there are signs of poor perfusion
- OR
- Deteriorating OI or OI >20
- OR
- Pre/post-ductal saturation difference of >10%
- For further guidance refer to separate PPHN Guideline.
- iNO may not provide the same benefits in infants with pulmonary hypertension where CDH is the cause of the pulmonary hypertension.

Extracorporeal Membrane Oxygenation (ECMO)

- The benefit of ECMO in infants with CDH remains unclear. Evidence suggests no clear survival benefit from its use and a possible increase in long-term disability in survivors. Even in immediate ECMO use (EXIT-to-ECMO) whilst infants are still on placental circulation, there is no evidence of improved outcomes.

- The usual indications and contraindications to the use of ECMO apply, including irreversible lung disease
- Criteria for early discussion with a local ECMO centre are:
 - Inability to maintain preductal saturations >85% or postductal saturations >70%
 - Increased PaCO₂ with pH <7.15 despite optimum ventilator management
 - PIP >28 cm H₂O or mean airway pressure (MAP) >17 cm H₂O to achieve saturation >85%
 - Metabolic acidosis with lactate >5 mmol/L and pH <7.15
 - OI ≥ 30
 - Systemic hypotension resistant to fluid and inotropic support resulting in urine output <0.5ml/kg/h for 12-24 hours

Surgical Repair

- The timing of optimum surgical repair is unknown; perform surgery electively when the infant is clinically stable.
- The routine use of a chest drain postoperatively is not recommended unless required in individual cases for symptomatic effusions.

Post-Operative Care

- Perform a post-operative chest x-ray to confirm ETT position, lung expansion and line positions (if present).
 - Note, the ipsilateral hemithorax will be air filled, this will gradually fill with fluid over the first few days post-surgery, drainage is rarely required.
- Do not aspirate or drain air or fluid without consultation with a consultant surgeon and neonatologist.
- Ventilation considerations are described above (Ventilation Management), in particular lower distending pressures results in improved respiratory function after surgical repair, possibly due to hypoplastic lungs in CDH being prone to overdistension.
- Assess carefully fluid balance; consider fluid restriction to 40-60ml/kg/d and diuretics if there is a persistent positive fluid balance.
- Start preventative anti-reflux therapy when enteral feeding is started.

Long-Term Follow-Up

- CDH survivors have multiple long-term medical problems, most commonly chronic lung disease, gastro-oesophageal reflux, poor postnatal growth and neurodevelopmental delay
- The following should be arranged pre discharge:
 - Neonatal follow up in outpatient clinic
 - Paediatric surgery clinic follow up
 - Paediatric respiratory referral
 - Audiology referral
 - Dietician as required
 - Speech and language as required
 - Cardiology follow up if persistent pulmonary hypertension
 - Palivizumab prophylaxis should be arranged