INPATIENT MANAGEMENT STANDARDS FOR CDH

Delivery Room Management

Principles

The main principle in the immediate management of a baby with CDH at birth is the early establishment of a definitive airway by intubation, avoiding mask ventilation. This should minimise gastric/bowel distension and prevent further deterioration due to expansion of the herniated structures. Early placement of a gastric tube is also essential.

The initial goals of ventilation should be to produce discernible chest movement and a stable, adequate heart rate. Oxygen saturation at this early stage is of lesser importance. Basic monitoring: heart rate, non-invasive blood pressure and pre-ductal saturation, should suffice in the delivery room and the baby should be moved to NICU as soon as possible. Establishment of invasive monitoring and central venous access should be deferred until the baby is in NICU. Aim for pre-ductal saturations of 80–95%, although lower saturations are acceptable if heart rate is stable and saturation gradually increasing. Peak ventilation pressure should be kept ≤25cmH_20, although higher pressures may be required briefly to achieve initial ventilation.

If there is evidence of poor perfusion or hypotension then initial treatment should be one or two boluses of 10ml/kg 0.9% saline aiming for a mean BP normal for gestational age.

For a baby born outwith one of the centres with paediatric surgery, discussion should take place with the specialist team as soon as possible and certainly within 2 hours of birth.

Initial contacts for the three surgical centres are:

Glasgow - On call Consultant Neonatologist or Consultant Neonatal Surgeon Switchboard 0141 201 0000, NICU sister's office 0141 201 0462

Edinburgh - On call Surgical Registrar or Consultant Surgeon via RHSC switchboard 0131 536 0000.

On Call Consultant Neonatologist via RIE switchboard 0131 536 1000

Aberdeen - On Call Consultant Neonatologist or Consultant Neonatal Surgeon via switchboard 0845 456 6000. Direct Number for Neonatal Unit 01224552602. North Regional Transport Team (to be used for this purpose only) 01224551767

Key Points for Delivery Room management

1. If the diagnosis of CDH has been made pre-natally, senior neonatology staff should be in attendance at the delivery.
2. For pre-natally diagnosed cases, endotracheal intubation should be carried out immediately, avoiding face mask ventilation. In the case of a baby with no pre-natal diagnosis but early respiratory distress, intubation should be carried out as soon as the diagnosis of CDH is suspected.
3. Pedicap or other capnography devices may not be reliable in confirming tube position.
4. Initial monitoring – heart rate, NIBP and pre-ductal SaO_2
5. Suggested initial ventilation pressures are 25/5cm H_2O, increasing peak pressure as required to achieve discernible visible chest wall movement and an adequate stable heart rate.
6. A large bore (10F) oro-gastric or naso-gastric tube should be passed, aspirated and left on free drainage.
7. Peripheral vascular access should be obtained
8. Sedate and paralyse
9. If poor perfusion or hypotension (below normal mean BP for gestation) consider up to two 10ml/kg boluses of 0.9% Saline.
10. Transfer to NICU

NICU/PICU management

1. Monitoring and vascular access

a. Pre- and post-ductal SaO_2

b. Invasive arterial blood pressure (It is not essential to have a "pre-ductal" arterial line. An umbilical artery catheter is satisfactory.)

c. Routine NICU intensive care level monitoring

d. Central venous access should be obtained preferably with a double-lumen umbilical venous catheter.

e. Chest X Ray should be performed as soon as possible to confirm the diagnosis, assess severity and confirm satisfactory placement of ET and NG tubes.

Echocardiography is valuable in guiding both the therapy of pulmonary hypertension and circulatory support as well as the diagnosis of associated cardiac anomalies.

2. Ventilation

a. Principles and ventilatory goals

CDH is associated with varying degrees of pulmonary hypoplasia. There is broad consensus that ventilation with high peak pressures or tidal volumes in an effort to achieve "normal" blood gases causes lung injury and a progressive decline in lung function. A strategy of "gentle ventilation" with permissive hypercapnia is standard practice. For conventional IMV, PIP should be kept ≤25cmH_2O and tidal volume below 5ml/kg to prevent lung injury.

The goals of ventilation in terms of blood gases and oxygen saturations should be set lower than normal. In the first 2 hours a pre-ductal saturation as low as 70% is acceptable as long as this is slowly improving and there is evidence of satisfactory tissue perfusion as evidenced by an arterial pH >7.2 (H^+ X)and a pCO_2 in the target range. Subsequently pre-ductal oxygen saturation should be between 85% and 95%. In some cases pre-ductal saturations as low as 80% may be accepted as long as there is evidence of adequate organ perfusion as indicated by an arterial pH >7.2, lactate <5mmol/l and a urine output greater than 1ml/kg/hr. Arterial pCO_2 should be between 6–8kPa or even higher as long as arterial pH and oxygenation are acceptable.

b. Conventional

PIP should be limited to 25cmH_2O with a PEEP of 2-5cmH_2O and rate adjusted to achieve a pCO_2 between 6-8kPa. If goals cannot be achieved within these parameters then other ventilation or treatment modalities should be considered.

c. HFOV

If CO_2 cannot be controlled at an acceptable level on conventional ventilation then HFOV should be considered. HFOV may also improve oxygenation, although there is a lack of evidence from randomised trials that it influences mortality in CDH.

Suggested initial settings are MAP 13-17cmH_2O, frequency 10Hz and Δp 30-50cmH_2O. Regular chest X-rays should be obtained to avoid over-inflation.

Key Points

1. Aim for pre-ductal oxygen saturation of 85-90%.
2. A pre-ductal saturation as low as 80% may be accepted if there is evidence of adequate organ perfusion/function.
3. Aim for arterial pCO_2 of 6-8kPa, although higher levels may be tolerated if pH/H^+ and oxygenation acceptable.
4. PIP on conventional ventilation should be kept ≤25cmH_2O.
5. If goals not achieved on conventional ventilation with PIP <25cm H_2O then consider HFOV, management of pulmonary hypertension or ECMO.

3. Pulmonary hypertension

In the lung of infants with CDH there are fewer divisions of the pulmonary arteries, resulting in a decreased cross sectional area of the pulmonary vascular bed. The walls of the pulmonary arteries also show increased wall thickness due to increased smooth muscle and increased reactivity. These changes frequently result in persistent pulmonary hypertension with hypoxaemia and right to left shunting. This may or may not manifest as a pre/post-ductal saturation difference depending on the level of the shunt but usually there will be a saturation difference of >10%.

Echocardiography is the best means of assessment of pulmonary circulation also giving information on right ventricular function/overload and left ventricular dysfunction. Left ventricular dysfunction either secondary to right ventricular overload or underdevelopment of the left ventricle is associated with a poor prognosis.

If the pre-ducal saturations are below 85% with evidence of poor organ perfusion, treatment of pulmonary hypertension should begin by optimising systemic blood pressure. Isotonic fluid boluses should be used initially as described in section 5 below, progressing to inotropic support if there is not an adequate response. The minimum target should be a mean BP normal for gestational age.

Persisting pulmonary hypertension should be treated with inhaled Nitric Oxide (iNO) starting at 10-20ppm. Response to treatment is best assessed by repeat echocardiography but a 10-20% reduction in the pre/post-ductal saturation difference and a similar improvement in pre-ductal saturation is good evidence of a satisfactory response. Whilst it is clear that iNO can improve physiological variables

there is no randomised controlled trial evidence of a reduction in mortality in infants with CDH.

In infants with systemic or suprasystemic pulmonary artery pressures, and a small or closed duct, the right ventricle may become overloaded with secondary left ventricular dysfunction and poor systemic perfusion. Shunting may be evident at the foramen ovale. In this circumstance, Prostaglandin E1 (Prostin) may be used to re-open the duct and offload the right ventricle. This should be guided by regular echocardiographic assessment.

Sildenafil has been used in the chronic treatment of pulmonary hypertension in the post operative phase in infants with CDH. There is currently insufficient evidence to support its use in the acute phase of management.

Key Points

1. Early echocardiography is highly recommended. In a clinically stable infant, an urgent echocardiography is unlikely to change management, but in an unstable infant, echocardiographic assessment of ventricular function and ductal patency is important to guide treatment of pulmonary hypertension.
2. Systemic mean blood pressure should be maintained at or above the level normal for gestational age.
3. iNO should be considered if there is hypoxaemia or poor perfusion associated with >10% pre/post-ductal SaO_2 difference or echocardiographic evidence of pulmonary hypertension.
4. If echocardiography shows a small/closed duct associated with suprasytemic pulmonary artery pressure, right ventricular overload, or right to left shunt at the foramen ovale, consider using prostaglandin E1.

4. Surfactant

There is some evidence to suggest that use of surfactant may be associated with a higher mortality and other adverse outcomes in infants with CDH. This may also be the case in preterm infants with CDH. The routine use of surfactant is not, therefore, recommended.

5. Haemodynamic support

Pulmonary hypertension is common in infants with CDH and may result in right to left shunting with tissue hypoxia and acidosis. Maintaining an adequate systemic blood pressure will help reduce shunting. If ventilatory goals are being achieved, a normal blood pressure for gestational age should be for the minimum target, although higher blood values may be required particularly if there is evidence of right to left shunting with associated low saturations.

Initial haemodynamic support is with fluid boluses, giving up to two 10ml/kg boluses of 0.9% saline. Subsequently inotropes (and possibly hydrocortisone) should be

used according to local preferences. Echocardiography is very useful in assessing the response to therapy and guiding the choice of inotrope/pressor.

6. **Sedation and paralysis**

   All infants should receive adequate sedation according to local protocols. Ideally a validated analgesia and sedation scoring system should be used. Paralysis is often required, at least until initial stability is achieved.

7. **Refer/Discuss with Specialist Centre**

   Babies born outwith one of the three specialist paediatric surgical centres should be discussed with the nearest specialist team as soon as possible after birth and certainly within the first two hours.

8. **Fluid balance**

   Fluid management should be as for any newborn receiving IV fluids, with additional boluses of saline as above if required for circulatory support.

   If a positive fluid balance occurs then diuretics should be considered aiming for a urine output of at least 1-2 ml/kg/h.

9. **Enteral feeding and Gastro-oesophageal reflux.**

   Conventionally, enteral feeding is delayed until after surgical repair of the defect. Gastro-oesophageal reflux is common. There should be a low threshold for starting anti-reflux medications if there are symptoms or signs suggestive of reflux. Early involvement of a dietician is advised if there are issues with weight gain and growth.

10. **ECMO**

    The role of ECMO in the management of babies with CDH remains unclear. In non-randomised trials improved survival has been reported. A systematic review of retrospective studies suggests that ECMO does increase survival but a meta-analysis of sub groups in randomised trials, although indicating an early survival benefit, does not show a long-term advantage for ECMO. Most ECMO centres report survival rates of 50% for this high-risk subgroup.

    Criteria for consideration of ECMO are controversial. Oxygenation index (OI) and/or alveolar-arterial O_2 difference (A-a DO_2) have been used, but absolute values are not widely accepted.

    ECMO should be considered if there is a failure to achieve therapeutic goals with optimal ventilatory and haemodynamic management. In order to avoid offering ECMO to an infant with pulmonary hypoplasia incompatible with long-term survival, it has been suggested that adequate oxygenation and ventilation should be demonstrated, at least for a short period. This is dependent on optimal management from delivery, and avoidance of acute lung injury.

    The optimal timing of surgical repair in relation to ECMO also remains unclear. In general there is a trend towards pre-operative stabilisation on ECMO with early

repair on ECMO, unless there is evidence that weaning from ECMO will be achieved to allow post-ECMO repair.

Early discussion with the ECMO centre is encouraged. Initial contact should be with the Consultant Neonatologist in RHSC, Glasgow (available through switchboard 0141 201 0000). Alternatively, a message can be left with the Ward manager on NICU (0141 201 0462) and a Consultant Neonatologist or ECMO surgeon will return your call.

Suggested criteria for consideration of ECMO

1. Inability to maintain pre-ductal saturations >85% despite optimal ventilation and management of pulmonary hypertension.
2. Arterial pCO_2 above the target range with respiratory acidosis (pH<7.15) despite optimal ventilation.
3. PIP consistently above 25cmH_2O (on CMV) to achieve ventilatory goals, or failure to improve following conversion to HFOV.
4. Inadequate tissue oxygen delivery as evidenced by metabolic acidosis pH<7.15 and lactate >5mmol/l.
5. Systemic hypotension, resistant to fluid and inotropes with a urine output <0.5 ml/kg/hr over 12-24 hrs.
6. A period of adequate oxygenation/ventilation should be demonstrated with optimal management (e.g. pre-ductal SpO_2>85%, pCO_2<8kPa)

Contraindications for ECMO support

1. Evidence of intraventricular haemorrhage >Grade I
2. Weight <2000g, or gestation <34 weeks
3. Ongoing bleeding/uncontrollable coagulopathy
4. Major associated co-morbidities (cardiac, chromosomal)

The Scottish Paediatric ECMO Centre would encourage discussion of all cases that meet the first 5 criteria above.

Surgery

1. Timing of surgery

Surgical repair of the diaphragmatic defect should be deferred until the baby is stable: mean BP normal for gestational age, pre-ductal SaO_2 within the target range on an FiO_2 <0.5, normal lactate and urine output >2ml/kg/h.

2. Surgical Technique

Standard repair is via an upper quadrant laparotomy. A patch may be required for large defects. There is insufficient evidence to make a recommendation as to patch material or configuration. Defect size and configuration should be recorded in a standard fashion as part of the MCN data set.

An alternative surgical approach is repair thoracoscopically. There are data on the feasibility of this approach but it is not yet clear whether it offers advantages over a laparotomy or if it is associated with increased risks due to the CO_2 pneumothorax or an increased risk of recurrence. At present it is recommended that thoracoscopic repair should only be undertaken by surgeons and anaesthetists with extensive experience of endoscopic surgery and in patients who are haemodynamically stable. Any patients who have thoracoscopic repair should be subject to close follow up.

Routine use of a chest drain following CDH repair, by either route, is not recommended.

References

1. Standardised postnatal management of infants with congenital diaphragmatic hernia in Europe: The CDH EURO Consortium consensus. Neonatology 2010;98:354-364
2. Thoracoscopic repair of congenital diaphragmatic hernia in neonates. NICE January 2011
3. Congenital diaphragmatic hernia: a systematic review and summary of best evidence practice strategies. Journal of Perinatology 2007;27:535-549
4. Mechanical ventilation strategies in the management of congenital diaphragmatic hernia. Seminars in Paediatric Surgery 2007;16;115-125