CONGENITAL DIAPHRAGMATIC HERNIA - POSTNATAL MANAGEMENT

- 86% of congenital diaphragmatic hernias (CDH) are left sided
- 40-50% have other congenital abnormalities, most importantly cardiac and cerebral defects
- Antenatal predictors of poor outcome are
  - Observed/Expected Lung Head Ratio (O/E LHR)\(^1\)
    - >45%  100% survival
    - 36-45%  ≈70% survival
    - 26-35%  ≈50% survival
    - 0-25%  ≈20% survival
  - L sided CDH with >20% of the liver in the thorax\(^2\)
- Overall survival is 80% or better in isolated CDH using lung protective ventilation +/- ECMO.
- Associated major cardiac anomaly decreases survival by about 50%
- Postnatal predictors of poor outcome are 5 minute Apgar score <7, birth weight <1500g, major heart anomaly, chromosomal anomaly, and severe pulmonary hypertension\(^5\)
- Discuss all cases with NICU and PICU prior to delivery and liaise closely post delivery
- OI = Oxygen Index = Mean Airway Pressure/ PaO\(_2\) (mmHg) x FiO\(_2\) (%). This should always be calculated using the preductal (right arm) PaO\(_2\). A postductal (umbilical or leg) PaO\(_2\) is not a valid indicator of disease severity. As a guide to indicate specific therapies, an OI>20 would suggest a trial of iNO and an OI>40 may be an indication for ECMO

Stabilisation/resuscitation at birth
- Intubation immediately after birth with no bag mask ventilation
- Pre-ductal oximetry
- Neopuff ventilation using goals as per ventilation section
- Stomach decompression with nasogastric tube
- Weigh before transfer to incubator
- Update parent(s) before leaving the room

Stabilisation in NICU
- Commence conventional ventilation as per ventilation section
- Measure pre and post ductal saturations
- Central venous access – double lumen umbilical venous catheter. An internal jugular (IJ) or femoral venous catheter may be inserted later in PICU. The right IJ will need to be cannulated if the patient needs ECMO
- A right radial/brachial/axillary arterial catheter is desirable to allow assessment of preductal blood gases. If this is not possible, a postductal arterial line (e.g. umbilical arterial catheter) and preductal saturation monitoring is acceptable
- Sedate and consider paralysis
- Maintain mean arterial pressure in normal range for gestation with fluid boluses +/- inotropes
- Obtain a chest X-Ray

Ventilation
- The aim is to achieve preductal SaO\(_2\) 80-95% in the delivery room and initial 1-2 hours of life. Lower saturations (70-80%) may be accepted over this time provided that they are improving without ventilator changes
- All hand bagging requires an in-line manometer or pressure controlled T-piece (e.g. Neopuff) to limit PIP ≤ 25cmH\(_2\)O
- Goals are:
Paediatric and Neonatal Intensive Care Units

Starship Children’s Hospital

Author: Buckley/Beca/Hegarty/Alsweiler
Page 2 of 4
Updated: 20/08/16

- Preductal saturations 80-95% (preferably 85-95%)
- Postductal saturations >70%
- PaCO₂ 6.7-9.3 kPa
- pH >7.2

- Pressure control ventilation should be used initially with the following strategy:
  - PIP ≤ 25 cmH₂O
  - PEEP 3-5 cmH₂O
  - Rate 40-60/min
  - Reduce FiO₂ if preductal saturations are >95%
  - Reduce rate or pressure if PaCO₂ < 6.7 kPa

- If oxygenation and/or ventilation goals cannot be met with a PIP ≤ 28 cmH₂O with conventional ventilation then HFOV may be used as a rescue therapy. A recent trial showed more pulmonary hypertension therapy, ECMO and longer length of stay for HFOV compared to conventional ventilation.

- With HFOV the goal is for the mean airway pressure to be ≤ 16 cmH₂O and no recruitment manoeuvres are to be used. Initial settings should be MAP 13-15 cmH₂O, f=10 Hz, delta P 30-40 (max 45). There should be frequent assessment with CXR (at least daily) to assess for overdistension (>8 ribs)

- There is no role for routine use of surfactant

Haemodynamic Monitoring and Management

- Haemodynamic goals are a normal heart rate and blood pressure for gestational age, a lactate <3 mmol/L and urine output >1 ml/kg/hr
- Ductal shunting and hypotension is treated with volume loading and inotropic support. It is usually because of right heart dysfunction
- Hypotension should be treated initially (in the first few hours) with a 10-20 ml/kg fluid bolus and this can be repeated once. If hypotension persists, vasoactive drugs should be used. Initially low dose adrenaline (0.03-0.05 mcg/kg/min) with noradrenaline if hypotension persists. Stress dose steroids (hydrocortisone 1 mg/kg 8 hrly) should be used if noradrenaline is required

Pulmonary Vascular Management

- Cardiac echo should be performed once stable after birth to (i) look for congenital heart disease (ii) assess right heart function and (iii) determine the degree of pulmonary hypertension (more or less than 2/3 systemic)
- Obtain a repeat cardiac echo if there is new or persistent cardiovascular instability to assess volume state, ventricular function, patency/size of the ductus, and pulmonary pressures
- Increased pulmonary vascular resistance (PVR) is almost universal and is the main determinant of survival. Right to left ductal shunting occurs when pulmonary artery pressure exceeds mean arterial pressure. This is not harmful as long as preductal saturations are maintained as these reflect cerebral oxygen delivery
- Hyperventilation should never be used to reduce PVR as it may damage the hypoplastic lungs
- Inhaled NO (iNO) should not be used routinely
- iNO should be tried if there is any of: (i) inability to maintain preductal saturations ≥85% (ii) OI >20 (iii) pre-post ductal saturation difference >10%
- iNO should be started at 20 ppm
- A response is indicated by (i) a 10-20% decrease in pre-post ductal saturation difference (ii) a 10-20% increase in PaO₂ or (iii) an improvement in mean blood pressure of 10% or more or a decrease in lactate
- If there is an insufficient response to iNO then other options include IV milrinone, IV sildenafil, IV prostacyclin, NG bosentan
- PGE₁ can be used to maintain ductal patency and prevent RV failure. It should be considered early if PA pressures are near systemic or suprasystemic, even if oxygenation is satisfactory.
New preductal desaturation in a previously stable baby may also indicate that the ductus has closed or become restrictive. An echo should be performed and PGE₁ started.

- Increasing the pH with sodium bicarbonate should only be used if there is pulmonary hypertension with haemodynamic compromise. If this is not present a pH as low as 7.20 is acceptable.

**Other measures**
- Muscle relaxants: These may be administered for initial stabilisation but do not need to be continued if the child is stable and movement does not interfere with oxygenation or increase ductal shunting. Generally with good prognostic features (e.g. O/E LHR >50%, liver down) muscle relaxants should not be necessary.
- NBM and IVN prior to repair.
- Head and renal ultrasound scan when stable.
- Chromosomal testing as appropriate.
- Group and Screen blood.

**Transport between NICU and PICU**
- This should occur at the soonest practical opportunity but is usually not urgent and can generally happen in daylight hours.
- The NICU team should perform transport, after consultation between the medical and nursing teams of NICU and PICU.
- The NICU team is responsible for management decisions until the patient’s care is handed over to the PICU team at the bedside in PICU.
- Handover will be consultant to consultant.
- The practicalities of the transport e.g. equipment, route taken, personnel, will be decided by the transporting team with an emphasis on patient safety. See “Transferring a Baby to PICU on VN500 – Oscillation Mode” in NICU guidelines.
- There will be the occasional patient who is too unstable to be transported and who may need to be placed on ECMO in NICU.

**On Arrival to PICU**
- Baby to remain on heat table/in incubator until handover complete.
- Request presence of PICU team members for single team handover.
- NICU Consultant gives SBAR summary followed by detailed systems handover.
- NICU Bedside Nurse completes handover.
- Combined team physical transfer of patient to PICU bed (unless have agreed continued use of NICU heat table in PICU) with tasks allocated to individuals and one Event Manager with oversight. Including transfer of syringes to PICU syringe drivers as necessary.
- Parent support person from NICU ensures parent(s) handed over to PICU team.

**ECMO**
- There is increasing use of ECMO in CDH (approximately 30% of all infants in many series) with good results. Patients who require high dose inotropes and/or cannot achieve acceptable preductal saturations with “gentle ventilation” should be considered for ECMO early. Decision will be influenced by O/E LHR and presence of associated anomalies/syndromes.
- Criteria for ECMO:
  - Inability to maintain preductal saturations >85%.
  - Requiring either PIP>28cmH₂O or MAP>16 cmH₂O to maintain preductal saturations >85%.
  - Oxygenation index (based on preductal sample) >40 for >3hr.
  - Respiratory acidosis with pH < 7.15 on maximum ventilation settings.
  - Inadequate tissue oxygen delivery – e.g. lactate > 5mmol/L.
Surgery

- Timing is contentious but it is an elective procedure. Surgery can lead to deterioration in thoracic compliance and increased PaCO₂.
- Surgery is generally performed when there is
  - Normal mean blood pressure for gestation
  - Preductal saturations of 85-95% on FiO₂ ≤50%
  - Normal lactate and urine output
- In patients with low PIP, low FiO₂ and minimal shunting, surgery can occur in the first 24-48 hours.
- Where possible, infants on HFOV should be transitioned to conventional ventilation before surgery.
- Patients who have bad prognostic features (e.g. >20% liver in the chest) and severe pulmonary HT may benefit from early surgery and ECMO afterward if necessary.
- Patients who require ECMO do better with early surgery either before or within 48 hours of commencing ECMO but need meticulous haemostasis to avoid major bleeding problems.
- In unstable patients surgery should be carried out in PICU.
- Pleural drains are not placed at the time of surgery. They are very rarely required after surgery for pleural effusions causing large mediastinal shift or pneumothorax.
- The principles of management after surgery are the same as before surgery.
- Enteral feeding should be started as soon as the postop ileus has resolved. Feed intolerance is common and feeds usually need to be increased in volume slowly.

References

5. Brindle et al, Pediatrics, 2014;134:2,e413-9