CONGENITAL DIAPHRAGMATIC HERNIA

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a congenital defect in the diaphragm (usually detected antenatally) resulting in herniation of abdominal contents into the thoracic cavity; associated with a high risk of mortality and morbidity. A combination of pulmonary hypoplasia and abnormal morphology of the pulmonary vasculature leads to severe respiratory insufficiency and increased risk of developing persistent pulmonary hypertension (PPHN)

RECOGNITION AND ASSESSMENT

Antenatal diagnosis

- Delivery to be planned at regional neonatal intensive care unit (NICU)
- Fetal medicine team and paediatric surgeon to provide antenatal counseling
- Neonatal team to meet parents before delivery
- Neonatal consultant, tier 2 staff and NICU nurse to attend delivery

Postnatal diagnosis

- In some babies the lesion develops later in gestation; these babies tend to have a better prognosis
- Postnatal presentation can be with clinical features ranging from inability to resuscitate baby at birth to incidental finding on chest X-ray

In cases diagnosed postnatally there may be early respiratory distress in association with a scaphoid abdomen and heart sounds shifted usually to the right. Mask inflation will often cause deterioration as air is delivered into herniated gut resulting in cardiorespiratory embarrassment

INVESTIGATIONS

- Pre and postductal SpO_2
- Chest and abdominal X-ray
- Arterial blood gas
- Echocardiogram

IMMEDIATE MANAGEMENT AT DELIVERY

Key principles

- Intubate all antenatally diagnosed babies promptly (intubation to be carried out by most experienced and reliable operator present)
- Optimise ETT position and size, aiming for little or no leak, with largest size tube feasible
- confirm tube position by end tidal CO_2 monitoring
- Do not give mask ventilation – will introduce air into the GI tract and compromise ventilation
- Maintain low peak pressure <25 cm H_2O and positive end expiratory pressures of 5 cm H_2O to avoid lung damage
- Avoid high airway pressures
- Establish adequate perfusion and oxygenation
- aim for preductal SpO_2 80–95% after first 10 min
- avoid hyperoxia, reduce FiO_2 when preductal saturation >95%
- Insert large gauge 8–10 Fr NGT
- aspirate at least every 5 min to decompress stomach until baby established on ventilation, then place on free drainage
- Check temperature before transfer to NNU, maintain normothermia
- Examine baby for other associated abnormalities e.g.:
- cardiac (present in 20%)
- trisomy 18/21
- urogenital
- musculoskeletal

MANAGEMENT ON NNU

Undertake management PROMPTLY
Babies with CDH fare better with minimal handling – handle baby as little and as gently as possible

- Weigh baby
- Start on conventional ventilation with low tidal volume strategy of 3–4 mL/kg
- Sedation: morphine 20 micrograms/kg/hr
- Umbilical venous and arterial catheters
  - to be sited by experienced operator (initial management is time critical)
  - if not possible to site umbilical arterial catheter (UAC), insert peripheral arterial line
- Monitor pre and postductal SpO_2
  - first 2 hr **only**: if pH >7.2, $PaCO_2$ <8.6 kPa and saturations improving, aim for preductal saturations of >70%
  - **after** first 2 hr: if lactate and pH acceptable (pH >7.2, lactate <5 mmol/L) on arterial blood gas, aim for preductal SpO_2 80–95% (UAC measures postductal PaO_2) and postductal SpO_2 >70%
  - an abnormal lactate is an indicator of poor perfusion and must be corrected before the interpretation of acceptable levels of SpO_2
- Maintain arterial blood pressure at normal level for gestational age
- Surfactant **NOT** routinely recommended. Only to be administered after discussion with regional centre as risk of over-distension and pneumothorax
- Cardiac echocardiogram (ideally within 6 hr of birth) to:
  - exclude associated congenital cardiac disease
  - assess right ventricular function
  - look for evidence of persistent pulmonary hypertension (see **Pulmonary hypertension** guideline)
  - identify patent ductus arteriosus and assess shunting (see **Patent ductus arteriosus** guideline)

Ventilation
Gentle conventional (see* Ventilation: conventional *guideline)
- Avoid peak pressures >25 cm H_2O, maintain PEEP 3–5 cm H_2O
  - if greater peak pressures required to maintain preductal SpO_2 >80% and postductal SpO_2 >70%, discuss HFOV with consultant
    - if HFOV not available discuss with specialist centre e.g. KIDS NTS/BWCH to expedite retrieval

HFOV [see* Ventilation: high frequency oscillatory (HFOV) *guideline]
- Initial setting:
  - MAP: 12 cm H_2O (do not increase >16 cm H_2O)
  - rate/frequency: 10 Hz, delta P 25
- Chest X-ray 1 hr after commencing HFOV
  - if >8 rib spaces visible, lungs are hyper-inflated – reduce MAP

Target O_2 saturations
- Aim for preductal SpO_2 of 80–95%
  - if MAP >12 cm H_2O and FiO_2 >0.6 to maintain preductal SpO_2 >80%, commence inhaled nitric oxide (iNO) at 20 ppm (see **Nitric oxide** guideline – KIDS NTS to commence nitric oxide if required)

Permissive hypercapnia
- If pH >7.2, lactate <5 and urine output >1 mL/kg/hr: target $PaCO_2$ 6.9–9.3 kPa

Systemic blood pressure support
- Invasive blood pressure monitoring required
- If preductal SpO_2 80–95%, aim for mean arterial blood pressure corresponding to gestation

- Maintenance fluid volume: 60 mL/kg/day
- Treat hypotension or poor tissue perfusion (rising lactate, urine output <1 mL/kg/hr) with fluid boluses sodium chloride 0.9% 10 mL/kg (maximum 30 mL/kg)
- If heart rate normal, urine output >1 mL/kg/hr, lactate <3 – do not give inotropes
- If persistent hypotension or hypoperfusion and difficulty maintaining preductal saturation 80–95%, give inotropes

Term babies

- Start adrenaline 100–1000 nanogram/kg/min as first line (starting dose usually 300 nanogram/kg/min)
- If right ventricular failure on echocardiogram discuss adrenaline with specialist centre e.g. KIDS NTS
- If right ventricular failure add milrinone 35–45 microgram/kg/hr (starting dose 35 micogram/kg/hr; **DO NOT** give loading dose) and noradrenaline base 20–100 nanogram/kg/min (starting dose 20 nanogram/kg/min)

Preterm babies

- Start dopamine 10 microgram/kg/min and increase to 20 microgram/kg/min
- if excessive tachycardia (heart rate >200 bpm) secondary to dopamine, discuss with specialist centre e.g. KIDS NTS and add noradrenaline
- Monitor lactate – rise in lactic acidosis suggests excessive vasoconstriction by inotropes

Metabolic acidosis

- Accept pH >7.2
- Review vasoconstrictor effects versus benefits of inotropes
- Correct metabolic acidosis with sodium bicarbonate 4.2%; give full correction over 6 hr titrating against pH/base excess every 1–2 hr

MANAGEMENT OF PPHN

- Anticipate PPHN in babies with CDH
- Monitor pre and postductal SpO_2
- Calculate oxygenation index (OI) (UAC is a measure of postductal saturation)
- if OI >20 and/or pre:postductal saturation difference >10% discuss with tertiary centre e.g. KIDS NTS to expedite retrieval
- Initiate trial of iNO, 20 ppm for 1 hr
- Magnesium sulfate ($MgSO_4$,) is an effective pulmonary vasodilator, commence an infusion of $MgSO_4$ to achieve serum (Mg) above the normal range (>1 mmol/L)
- **can give rise to profound systemic hypotension**, use only in conjunction with active management of systemic blood pressure support
- Maintain arterial PaO_2 8–10 kPa
- See **Pulmonary hypertension** guideline

GENERAL SUPPORT

- Fluid: restrict to 60 mL/kg/day
- Keep large bore NGT on free drainage and regular aspiration, and nil-by-mouth. Buccal colostrum can be given if available
- Commence parenteral nutrition
- Send blood culture and commence first line antibiotics
- Send clotting screen and correct any abnormalities
- Correct hypocalcaemia
- If antenatal diagnosis of a duct dependent congenital cardiac lesion, or any uncertainty about the presence of cardiac anomaly, commence dinoprostone (prostaglandin E2) 5 nanogram/kg/min
- Maintain magnesium >1 mmol/L
- Maintain normothermia
- Monitor for pneumothorax. See **Chest drain insertion – Seldinger technique** and **Chest drain insertion – Traditional** guidelines
- Crossmatch 1 unit of blood

- Cranial ultrasound scan
- Send blood for chromosomes with parental consent (if not done antenatally)
- Sedation: morphine 10–20 microgram/kg/hr, but avoid deep sedation
- Avoid neuromuscular blocking agents – use is associated with hypoxaemia
- Minimal handling, developmental care with swaddling/cocooning
- Keep area around baby quiet and lights dimmed

COMMUNICATION WITH SPECIALIST CENTRE

- Neonatal consultant to inform planned paediatric specialist centre e.g. Birmingham Children's Hospital/Birmingham Women's Hospital once baby stabilised. This will require conference call with referring consultant, on-call surgeon at specialist centre, neonatologist, PICU intensivist and transport consultant e.g. KIDS NTS, to discuss urgency of transfer and ongoing management
- Undertake transport of babies for surgery only when:
- mean arterial blood pressure normal for gestation
- lactate <3 mmol/L and urine output >1 mL/kg/hr
- ventilation reduced to low pressure settings
- FiO_2 on conventional ventilation 0.5, with preductal saturations 85–95%
- baby fit for surgery and stable for ≥24 hr (may take ≥3–10 days)

EXTRACORPOREAL LIFE SUPPORT (ECLS)

- See **Pulmonary hypertension** guideline
- If ECLS considered refer to specialist centre (e.g. via KIDS NTS team)

USEFUL INFORMATION

- https://bwc.nhs.uk/paediatric-surgery-treatments
- https://kids.bwc.nhs.uk/
- https://www.e-lfh.org.uk/programmes/paediatric-surgery/