Trust Guideline for the Management of Congenital Diaphragmatic Hernia (CDH) in Neonates

A clinical guideline recommended

For use in:	Children's services - Foetal Medicine
By:	Children's healthcare providers in the above areas (All staff in the named area)
For:	Neonatal period
Division responsible for document:	Women and Children's – Division 3
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This guideline has been approved by the Trust's Clinical Guidelines Assessment Panel as an aid to the diagnosis and management of relevant patients and clinical circumstances. Not every patient or situation fits neatly into a standard guideline scenario and the guideline must be interpreted and applied in practice in the light of prevailing clinical circumstances, the diagnostic and treatment options available and the professional judgement, knowledge and expertise of relevant clinicians. It is advised that the rationale for any departure from relevant guidance should be documented in the patient's case notes. The Trust's guidelines are made publicly available as part of the collective endeavour to continuously improve the quality of healthcare through sharing medical experience and knowledge. The Trust accepts no responsibility for any misunderstanding or misapplication of this document.

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Version and Document Control:

Version Number	Date of Update	Change Description	Author
1	25/10/2017	New document	Miss Alexandra Scarlett Dr Rahul Roy Mr Ashish Minocha
1.1	13/05/2021	Reviewed, no clinical changes	Miss Alexandra Scarlett Dr Rahul Roy Mr Ashish Minocha Dr Lee Smith

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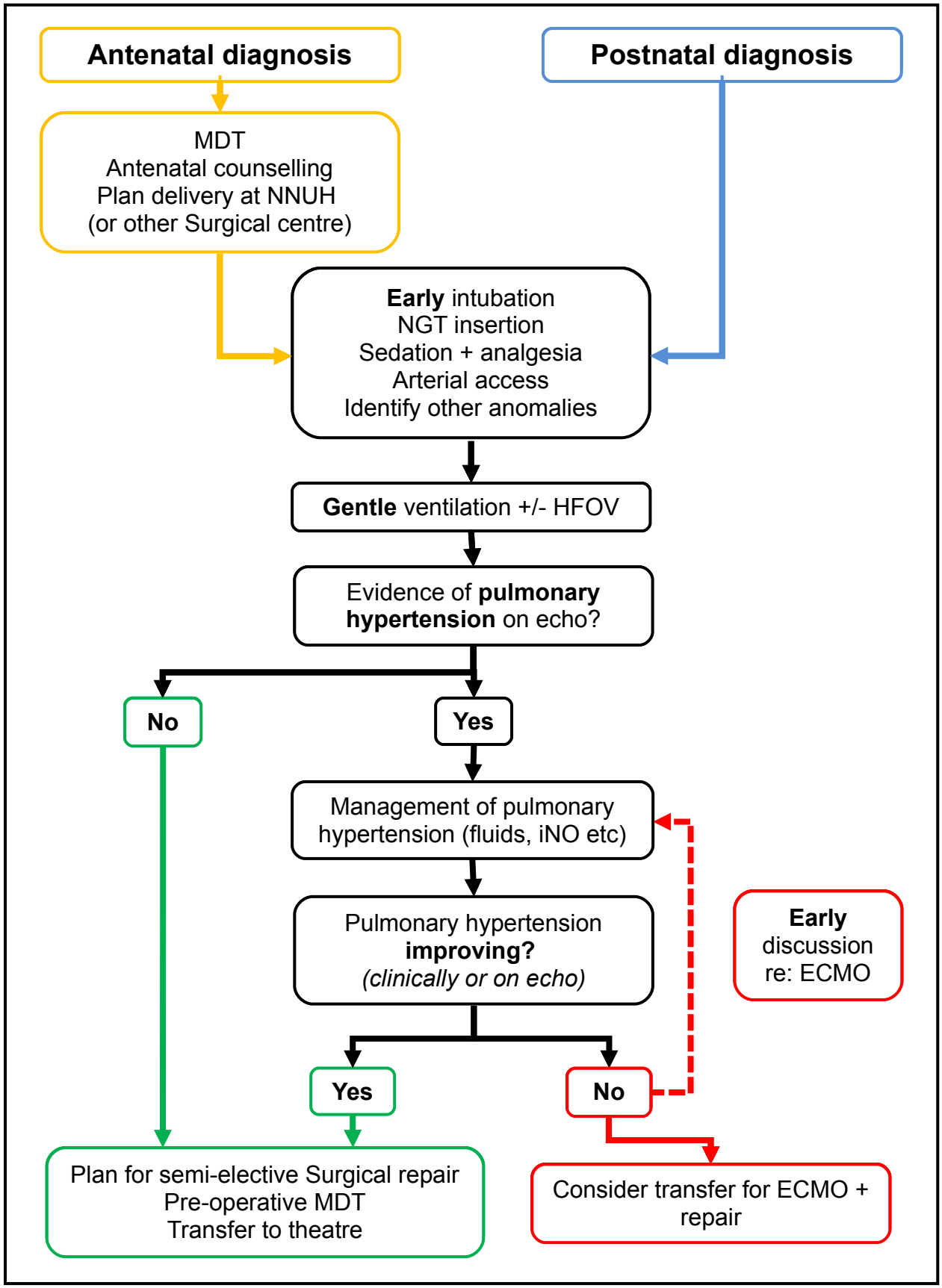
Quick reference

Management of Neonates with Congenital Diaphragmatic Hernia



Abbreviations

CDH Congenital diaphragmatic hernia

CPAP Continuous positive airway pressure

CXR Chest X-ray

DIC Disseminated intravascular coagulation

Echo Echocardiogram

ECMO Extra-corporal membrane oxygenation

ETT Endotracheal tube

FBC Full blood count (blood test)

HFOV High frequency oscillation ventilation

HRFR High risk feeding regimen

iNO Inhaled nitric oxide

IV Intravenous

MDT Multidisciplinary team

NICU Neonatal intensive care unit

NGT Nasogastric tube

OI Oxygenation Index

$$\text{Oxygenation Index (OI)} = \frac{\text{Mean Airway Pressure (cm } H_2O) \text{ x } FiO_2 \text{ x } 100}{PaO_2 \text{ (mmHg)}}$$

To convert from KiloPascals (KPa) to millmetres of Mercury (mmHg), multiply KPa value by 7.5

PEEP Positive end expiratory pressure

PIP Peak inspiratory pressure

PPHN Persistent pulmonary hypertension of the newborn

RR Respiratory rate

SALT Speech and language team

SDHCN Scottish Diaphragmatic Hernia Clinical Network

U+E Urea and electrolytes (blood test)

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Objective

The diagnosis and management of congenital diaphragmatic hernia (CDH) remains a complex and evolving process. The NNUH Trust Guideline aims to standardise care across the department; with a focus on reducing acute lung injury, appropriate consideration for use of ECMO and ultimately a timely and safe surgical repair.

Background

CDH is a multi-factorial, life-threatening, congenital malformation of the diaphragm. The diaphragmatic defect (most commonly posterolateral) allows an antenatal herniation of abdominal viscera into the thoracic cavity. The subsequent pressure affects lung growth and development bilaterally, causing lung hypoplasia. CDH incidence is 1 in 2000-3000 live births and predominantly (80%) affects the left side of the diaphragm. In 30% of cases CDH is associated with other congenital anomalies, (predominantly cardiac). CDH may also occur as part of a syndrome (eg Fryn's) or underlying chromosomal abnormality (eg. Trisomy 13 or 18). This association is up to 50% in the foetal population, but the anomalies often cause foetal demise. Antenatal scanning leads to a pre-natal diagnosis in 60-75% of cases allowing for appropriate counselling and planning.

Over the past few decades changes in ventilation management and delayed surgery have led to an improvement in survival rates. However overall survival remains 60%-70% (up to 90% with isolated CDH); lung hypoplasia, combined with pulmonary hypertension, is the leading cause for both morbidity and mortality.

Rationale

There is large variation in practice between, and within, departments. The formation of a standardised department guideline has been associated with increased rates of survival in this population; even the discussion improved care. The Perinatal Confidential Enquiry (MBRACE-UK 2014) found that having a clear pathway from diagnosis to follow up was an indicator of good or excellent care. The 2015 update of the **CDH EURO Consortium Consensus by *Snoek et al*** has been reviewed and incorporated into this guideline.

Key points

1) **Antenatal counselling** where possible and delivery at surgical centre
2) **Senior Neonatal team** should be present at delivery and NICU consultant informed
3) **Avoid face mask ventilation** and **intubate immediately**; if postnatally diagnosed then baby should be intubated as soon as CDH confirmed
4) **Gentle ventilation** with permissive hypercapnia, if signs of good tissue perfusion
5) **Minimise handling** to reduce PPHN crises
6) **Consider HFOV** if parameters not improving on conventional ventilation
7) **Consider ECMO** if pulmonary hypertension not improving on HFOV
8) **Delay surgery** until baby stable, after discussion with appropriate MDT

Antenatal management

Approximately 60% of CDH are diagnosed antenatally; further USS should be performed to assess the location of the defect, the observed/expected lung-to-head ratio (O/E LHR), the position of the liver, and to rule out other anomalies. Parents should have the opportunity to have **joint** antenatal counselling with the Neonatal and Paediatric Surgical team, in addition to sessions with Foetal medicine. There should be documented multi-disciplinary meetings regarding timing of delivery and plans to deliver in NNUH (or another Surgical centre). In the absence of other indications, planned delivery should not normally be before 39 weeks. There is no evidence that Caesarean section is beneficial.

Once baby is delivered and stable the On-call Paediatric Surgery SpR should be informed (on #1047).

Delivery Room management

Pre-operative management of babies with CDH will be led by the NICU team with close liaison from the Surgical team regarding timing of surgery.

If CDH is suspected antenatally:

- **Immediate intubation** and ventilation where possible (avoiding use of face mask ventilation, to minimise gastric distension).
- No routine use of **surfactant** in term infants.
- **Ventilation** with PIP as low as possible, aim for <25cmH_20.
- **Routine NICU monitoring** with pre-ductal SaO_2 monitoring aiming SaO_2 80-95%.
- **Place an 8Fr NGT** (or as large as possible), aspirate and leave on free drainage.
- **Peripheral IV access** consider bolus if signs of poor perfusion.
- **Early sedation** and **analgesia.**
- **Transfer** to NICU once stable.

Cases with a **postnatal diagnosis** often present within 24 hours, with respiratory distress of the newborn, and a suggestive CXR; classically these babies may also have a scaphoid abdomen and displaced heart sounds.

Once a CDH diagnosis is suspected these babies should be treated just as those diagnosed antenatally, with **early intubation**. Differential diagnoses include macrocystic congenital lung lesions or diaphragmatic eventration.

Further monitoring and early investigations

- Pre- and post-ductal saturation monitoring.
- Umbilical arterial catheter or other arterial line (consider right radial line first).
- Umbilical venous catheter or other central IV access.
- Catheter if necessary for monitoring urine output.
- CXR to confirm line/ETT position and confirm diagnosis.

- Echocardiogram to assess for structural cardiac anomalies, ventricular function and degree of pulmonary hypertension within first 24 hours after birth.

Ventilation

After swift intubation to minimise any further cardiac embarrassment, the initial goals of ventilation are to achieve visible chest movement and an appropriate heart rate. The general consensus is to avoid iatrogenic barotrauma by adhering to a strategy of 'gentle ventilation.' This is accomplished with the following:

- Aim to achieve pre-ductal SaO_2 85-95%, post-ductal >70%.

  In the first 2 hours pre-ductal SaO_2 >70% are acceptable **IF:**

  - *Pre-ductal SaO_2 is slowly improving.*
  - *Evidence of good organ perfusion (eg. pH >7.2).*
  - *Adequate ventilation (eg. $PaCO_2$ <8.6kPa).*
- Target $PaCO_2$ 7-9kPa (permissive hypercapnia).
- Initial ventilator settings of PIP <25cmH_2O, PEEP 3-5cmH_2O, RR 40-60bpm.

In individual cases pre-ductal SaO_2 down to 80% may be accepted, providing organs are well perfused, as indicated by a pH >7.2, urinary output >1ml/kg/hr and lactate levels <5mmol/L; post-ductal saturations should remain above 70%.

HFOV should be considered as rescue therapy if the CO_2 clearance and oxygenation remain inadequate (with PIP >28cmH_2O) with a CXR used to assess chest expansion. Although the recent randomised control trial demonstrated no difference in mortality using HFOV, there was a significant reduction in need for ECMO and time on the ventilator. If treatment goals continue to be 'not met' despite maximal medical management then early consideration of ECMO is recommended (*see below*).

Haemodynamic support

Optimising systemic blood pressure will help alleviate the processes secondary to PPHN, however a delicate balance is needed to prevent overload and its sequelae. Close beat-to-beat monitoring is required via an arterial line, aiming for mean arterial pressure equivalent to gestational age. Initial management may involve haemodynamic support with saline boluses (2x10-20ml/kg).

If symptoms of poor perfusion or hypotension with pre-ductal saturations <80% then echocardiogram should be performed.

Successive inotropic therapies may be obligatory to maintain adequate visceral perfusion. Please see NNUH Neonatal Hypotension guideline (Trustdocs ID: 7561) for more information.

Milrinone (a selective phosphodiesterase III inhibitor with both inotropic and lusitropic action) can be used to good effect to improve cardiac function by pulmonary and systemic vasodilation, and reducing afterload.

Management of pulmonary hypertension

The cross-sectional area of the pulmonary vascular bed of a baby with CDH is significantly reduced, due to fewer divisions of the pulmonary arteries. This, coupled with an increased pulmonary vascular wall thickness, results in pulmonary hypertension with hypoxaemia and a RIGHT-to-LEFT shunt. The aims of management are to normalise arterial oxygen concentration and optimise tissue oxygenation. Ventilation measures to achieve this are mentioned above, however optimising the systemic blood pressure will help alleviate the shunting and ensuing hypoxia. Serial echocardiogram can be utilised to assess changes in pulmonary arterial pressures, with the first echo ideally performed within first 24 hours.

Inhaled Nitric Oxide (iNO) acts as a vasodilator to improve oxygenation. If there is evidence of extrapulmonary right to left shunting and an OI >20, and/or pre-/postductal SaO_2 difference >10% then iNO can be used starting at 20ppm for 1 hour. A good response to iNO is indicated by:

- Decline of 20% in pre/post-ductal saturation difference, or;
- Increase of PaO_2 by >3kPa, or;
- 20% decrease of oxygenation index.

iNO should be stopped if no response seen after an hour. There is currently no evidence to suggest iNO reduces the need for ECMO or mortality.

Prostin (prostaglandin E_1) may be considered in babies with suprasystemic pulmonary artery pressure and a closed or narrow ductus ateriosus to ameliorate impending right ventricular failure after discussion with ECMO team.

Sildenafil can be considered in the management of severe pulmonary hypertension.

Fluids and feeding

Babies should remain *nil by mouth* until after the defect repair (then as per operative plan), therefore all fluid and nutritional requirements will be administered parenterally. Fluid restriction and diuretics can be utilised to maintain appropriate fluid balance, aiming for a urine output of 1-2ml/kg/hr.

Sedation vs paralysis

Ensuring adequate analgesia and sedation is vital in early CDH management; pulmonary hypertensive crises may occur with handling, suctioning and other more invasive manoeuvres. In order to mitigate these episodes experienced nursing care and a ‘minimal handling’ policy are required. Sedation and analgesia are commonly achieved using morphine and midazolam.

Avoid paralysis if possible, but use of appropriate muscle relaxant can aid ventilation. Paralysis reduces swallowing and further gaseous distension and allows optimisation of ventilation efficacy. However it may lead to increased atelectasis, oedema and a ventilation:perfusion mismatch.

Other medications

- ***Empirical antibiotic therapy*** – to continue until cultures clear.

- ***Surfactant*** – no evidence of benefit in **term** CDH and may cause over-distension of aerated lung areas and possible pneumothorax (*should still be prescribed for preterm infants as per local guideline*).
- ***Anti-reflux therapy*** – low threshold for starting therapy in the presence of reflux symptoms.

Extra-corporal membrane oxygenation (ECMO)

ECMO is a form of cardiopulmonary bypass used in neonates in severe respiratory or circulatory failure. Its role within the purview of treating refractive pulmonary hypertension in CDH remains unclear, with only non randomised studies demonstrating improved survival. Most ECMO centres anecdotally report survival ~50% in this high risk sub-group of CDH patients. However it is not without complication and involves transfer to one of three centres in the UK (Glasgow, Leicester or GOSH).

The criteria for transfer for ECMO are not absolute. However, failure of management of PPHN with HFOV and maximal inotropic support, with poor oxygenation and CO_2 clearance is generally accepted as a principle. An OI of >40 is often quoted as a benchmark but is not widely accepted.

Consider ECMO when:

- Inability to maintain pre-ductal SaO_2 >85% or post-ductal >70%.
- Respiratory acidosis (pH<7.15 and raised $PaCO_2$) despite ventilatory optimisation.
- Requiring a PIP >28cmH_2O or MAP >17cmH_2O to achieve SaO_2 >85%.
- Evidence of poor tissue perfusion (lactate ≥5, pH <7.15).
- Systemic hypotension resistant to fluid and inotropic support (ie urine output <0.5ml/kg/hr for 12-24hrs).
- OI ≥40 present for at least 3 hours.

Contraindications for ECMO are <34 weeks gestation, <2kg weight, lethal congenital anomalies, significant intraventricular haemorrhage (>Grade 1) and DIC. All ECMO centres advocate early discussion regarding potential cases.

Surgery

Delayed surgical repair of CDH is widely accepted to improve survival. Most evidence suggests awaiting clinical signs of improving pulmonary hypertension prior to considering surgery. Ideally the baby should have an appropriate for gestation mean arterial pressure off inotropes, with a lactate <3mmol/L and urine output >1ml/kg/hr. There should be a documented pre-operative discussion between the Neonatal, Paediatric surgical and Paediatric anaesthetic consultants including review of all recent bloods (FBC, U+E, clotting screen and crossmatch).

The standard open procedure involves the following steps:

- Subcostal laparotomy.
- Resect hernial sac if present.
- Assess muscle/defect and the need for patch.

- Consider risk of compartment syndrome.
- No routine use of chest drain.
- Documentation of:

  Size and type of defect (see Appendix 1).

  Hernial contents.

  Type of repair.

  Clear post operative plan including ventilation, need for paralysis and antibiotic course.

Principles of post-operative care

Please refer to operative note for patient specific plans – may deviate from below:

- Pain control – with paracetamol/morphine +/- epidural in lower risk babies.
- Continue IV antibiotics for 48 hours and review with surgeons and cultures.
- Consider fluid restriction depending on volume of intra-operative fluid given.
- Consider starting on HRFR once NGT aspirates have reduced/cleared.
- Continue PN until established enteral feeding.
- Start anti-reflux medication.
- Consider further echo – guided as per pre-operative findings.
- Important to carry on any drugs started by ECMO centre such as sildenafil.

Factors for discharge

- Parental Resus training.
- Feeding established (NGT/bottle/breast) and parents trained (for NGT feeds).
- Adequate growth on current feeding plan (with dietician plan in place).
- SALT assessment as required.
- Home oxygen requirement assessment as required.
- Encourage parents to stop smoking – inform GP (for further support).
- Inform the Respiratory team of discharge.
- Contact with local paediatrician / Health visitor / Social worker as required.
- Assess for Palivizumab criteria / other routine vaccinations.
- Confirm plan regarding medications +/- follow up from ECMO team.

Follow-up

Infants surviving post CDH repair may experience significant pulmonary, gastrointestinal, cardiac, and neurological sequelae (especially cognitive developmental delay). Therefore a structured, multidisciplinary follow-up may be necessary for more complex patients, initially with the Neonatal and Paediatric Surgery teams. On discharge from the Neonatal clinic the patients should be referred to the Paediatric Respiratory team. However this may occur earlier if there are significant respiratory concerns.

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Audit of Guideline – to be performed in 2-3 years (prior to guideline review).

Audit standards

1) All patients suspected of having an antenatal diagnosis of CDH should have joint counselling from Neonatal and Surgical teams. [100%]
2) Documentation of pre-operative MDT should occur between neonatologist, surgeon and paediatric anaesthetist. [100%]
3) Documentation of rationale for mode of ventilation if PIP >25. [100%]
4) Documentation of CDH defect grade should be made in the surgical note. [100%]
5) All patients should have Medical and Surgical follow-up. [100%]

The audit results will be sent to Department Audit lead (currently Mr Richard England), who will ensure that these are discussed at relevant governance meetings to review the results and make recommendations for further action.

Reference Guidelines

Standardized Postnatal Management of Infants with Congenital Diaphragmatic Hernia in Europe: The CDH EURO Consortium Consensus – 2015 Update. *Snoek AG et al. Neonatology. 2016;110(1):66-74*

Inpatient management standards for CDH – Scottish Diaphragmatic Hernia Clinical Network – last reviewed December 2013
(http://www.sdhcn.scot.nhs.uk/publications/sdhcn-guidelines/)

Fetal Anomaly Scan Patient Information leaflet for CDH – July 2017, Public Health England's Fetal Anomaly Screening Programme (FASP) and British Association of Perinatal Medicine (BAPM)
(Fetal Anomaly Scan Patient Information leaflet)

Development and consultation process undertaken before registration and dissemination

The authors listed above drafted this document on behalf of NICU/Paediatric Surgery Consultants who has agreed the final content.

This version has been endorsed by the Clinical Guidelines Assessment Panel (CGAP)

Distribution List

Trust intranet

Children's services

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Foetal Medicine

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14. Currently *'in press'* for BAPM - Principles of Neonatal Surgery involvement in the management of congenital diaphragmatic hernia (G Walker)
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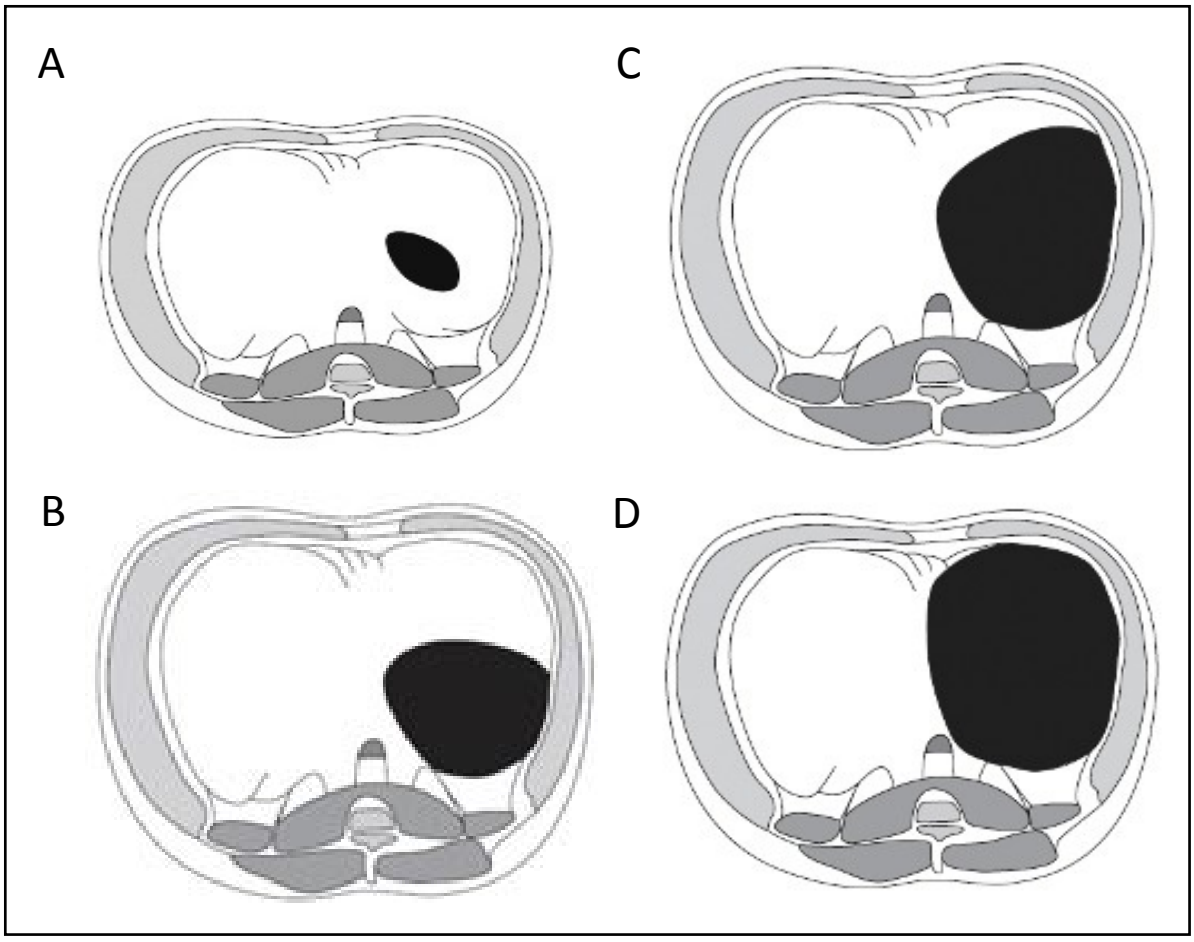
Appendix

Appendix 1 – Hernial defect grading



Grading of diaphragmatic hernia defect size at time of surgery. Reproduced from Lally *et al* (CDH study group).

Appendix 2 – for reference purposes only

Recommendation	Timing					
	Before discharge	1-3 months	4-6 months	9-12 months	15-18 months	Annually thereafter
General assessment and growth	√	√	√	√		√
Chest radiograph	√	If clinical indication	If clinical indication	√	If clinical indication	√
Pulmonary function testing			√	√		√
Childhood immunisations	Routine as indicated	Routine as indicated	Routine as indicated	Routine as indicated	Routine as indicated	Routine as indicated
RSV prophylaxis	√♦	√♦	√♦	√♦		
Influenza prophylaxis				√	√	√
Cardiology follow-up and echocardiogram	√	√♥	√♥	√♥	√♥	√♥
Cranial MRI	If clinical indication	If clinical indication	If clinical indication	If clinical indication	If clinical indication	If clinical indication
Hearing evaluation	√		√			√♦
Developmental screening assessment	√		√	√	√	√
Formal neuro-developmental evaluation						√ Bayley at 3 years
Upper GI investigations	If clinical indication	If clinical indication	If clinical indication	If clinical indication	If clinical indication	If clinical indication
Scoliosis and chest wall deformity screening				√		√

Recommended schedule of follow-up for infants/children with CDH – reproduced from Liddell et al; used within SDHCN, adapted from American Academy of Pediatrics recommendations
Key: ♦ - during first winter ♥ – evidence of PHT