Embrace Stabilisation and Transfer of Infants with Congenital Diaphragmatic Hernia

Reference:	1753v2
Written by:	Dr Chris Vas, Dr Cath Harrison
Peer reviewer:	Dr Jess Oldfield
Approved:	October 2021
Review Due:	October 2024
Embrace review due:	October 2022

Purpose

Assist Embrace clinicians in stabilising and transferring infants born with congenital diaphragmatic hernia outside of a tertiary neonatal unit. Guidance is also provided on the transfer from the Jessop Wing to Sheffield Children's Hospital.

Intended Audience

Embrace team (Nurse, ANPs and Medics)

Table of Contents

Glossary of abbreviations

ECMO	**Extra Corporeal Membrane Oxygenation**
PIP	**Peak Inspiratory Pressure**
CXR	**Chest x-ray**
UAC	**Umbilical arterial catheter**
UVC	**Umbilical venous catheter**
CRT	**Capillary refill time**
LGI	**Leeds General Infirmary**
O.I.	**Oxygenation Index**

1.1 Introduction

Congenital diaphragmatic hernia (CDH) is a defect in the diaphragm allowing abdominal contents to herniate into the chest. Most defects are isolated but it can be associated with congenital cardiac lesions and can be present with syndromes such as Downs, Edward's and Patau's. CDH has a high mortality rate[(1)] with the major pathophysiology being pulmonary hypoplasia and pulmonary hypertension.

1.2 Referral

CDH is usually antenatally diagnosed and delivery should be planned at a tertiary neonatal centre used to managing CDH [(2,3)]. These are:

- Leeds General Infirmary
- Jessop Wing, Sheffield

Infants born outside of these units with CDH need stabilisation and transfer via Embrace.

Infants at the Jessop Wing will need transferring via Embrace to Sheffield Children's Hospital for surgical repair once clinically indicated. These infants may then stay at Sheffield Children's Hospital or be transferred back to the Jessop Wing for ongoing care. **These infants need to be transferred to PICU the day before surgery.**

Infants with CDH sometimes require ECMO. Referral to the ECMO team may be undertaken by the tertiary unit directly or via Embrace.

1.3 Equipment

In addition to the normal equipment for transfer, the following are required:

- Nitric oxide tray and circuit
- Spare nitric cylinder

1.4 Risks and Precautions

- Consultant-led transfer if born outside tertiary centre or acutely unstable
- Stable infants on the unit may poorly tolerate transfer and can quickly deteriorate. Ensure adequate sedation and paralysis.

2.1 Acute stabilisation

Advice for Referring Units:

1. On delivery suite:

Known antenatal diagnosis but delivered outside specialist unit:

- Intubate immediately after delivery. Consider using IM paralysis (atracurium or pancuronium). Avoid bag and mask ventilation
- Ventilation: Start with pressures of 30/5 for 5 inflation breaths via endotracheal tube then reduce PIP to 20-25 ensuring chest movement. Start at rate of 60 breaths per minute. Obtain urgent CXR

- Oxygenation: Use 100% oxygen. Monitor pre-ductal SpO_2 and aim between 80-95%
- Insert an NG tube and put on free drainage. The NG needs to be sufficiently in the stomach so should be seen curling down and up on CXR

After moving to neonatal unit follow section 2.2 guidance

2. On neonatal unit, for postnatally diagnosed CDH:
   - Early senior assessment for signs of respiratory distress
   - Some infants with a postnatal diagnosis of CDH may have no signs of respiratory distress, as the diaphragmatic defect may have developed late on in pregnancy, and the lungs have been able to grow and develop as normal. They may be able to be transferred to a surgical centre with no respiratory support.
   - If there are signs of respiratory distress, follow the stabilisation as for on delivery suite.

2.2 Pre-departure Stabilisation

Ventilation:

- A lung protection strategy should be used to "gently" ventilate. The aim of ventilation is to avoid lung injury with acceptable ventilation and oxygenation. Initial PIP should be set between 20-25 with a rate of 40-60 to avoid barotrauma [3,7].
- Aim pre-ductal SpO_2 80-95% and post-ductal >70%. Consider weaning O_2 if SpO_2 >95%.
- Allow permissive hypercapnia, keeping pH>7.2 if infant needing higher pressures.
- In mild lung disease, allow normal pH and CO_2 to try to avoid pulmonary hypertension.
- No role for routine surfactant.

Cardiovascular:

- Intravenous access: Place peripheral cannula and send bloods. Consider sending for karyotype if not performed antenatally.
- Place UAC and double lumen UVC
- Commence Fluids: 10% glucose at 60mls/kg/day
- Aim for a normal blood pressure for gestation [3, 5]
- Assess perfusion with heart rate, CRT, urine output and lactate
- If hypotensive with poor perfusion or low SpO_2 then cautiously give 0.9% sodium chloride boluses, up to 30mls/kg
- If inotropic support is required then ideally this should be given via central access
- Inotrope choice will be guided by local considerations and advice from Tertiary Specialists [8,9]
- If a third line inotrope is required then this should be discussed via Embrace for consultant advice. Hydrocortisone, noradrenaline and/or adrenaline may be recommended
- See Embrace guideline for use of inotropes in Neonatal patients [9]

Pulmonary Hypertension:

- Consider pulmonary hypertension if pre and post ductal SpO_2 are both below 85% or there is a gap of more than 10% between the SpO_2
- Aim for mean BP > 45 mmHg to optimise systemic circulation
- Start nitric oxide if oxygenation index (O.I.) is greater than 20

O.I. = FiO_2 x Mean Airway pressure (cmH20)x100/PaO_2 (mmHg)
To convert kPa to mmHg multiply by 7.5

Sedation;

- Ensure adequate sedation. Start morphine at 20 micrograms/kg/hour and consider midazolam infusion
- Muscle relaxant for transfer is recommended. An atracurium infusion should be started at 0.5 mg/kg/hour and increased as needed

2.3 Indications for ECMO[3,4]

- Inability to keep pre-ductal SpO_2 above 85%
- OI>40 or OI >25 with inhaled nitric oxide
- PIP >28 and SpO_2 <85%

The role of ECMO is still unclear [6].
Infants with CDH will usually be transferred to a local tertiary unit for stabilisation before the consideration of ECMO, but early discussion with the ECMO team may be helpful. Infants at LGI or Jessop Wing can be referred for ECMO via Embrace if indicated with involvement from Leeds PICU for ECMO support.

3. References

1. Van den Hout L, Sluiter I, Gischler S, et al: Can we improve outcome of congenital diaphragmatic hernia? Pediatr Surg Int 2009;25:733.
2. Grushka JR, Laberge JM, Puligandla P,Skarsgard ED: Effect of hospital case volume on outcome in congenital diaphragmatic hernia: the experience of the Canadian Pediatric Surgery Network. J Pediatr Surg 2009;44:873.
3. National Care Principles for the Management of Congenital Diaphragmatic Hernia (2018). A BAPM Framework for Practice
4. Reiss I, Schaible T, van den Hout, L et al. Standardized Postnatal Management of Infants with Congenital Diaphragmatic Hernia in Europe: The CDH EURO Consortium Consensus. Neonatology 2010;98:354–364
5. Mugford M, Elbourne D, Field D: Extracorporeal membrane oxygenation for severe respiratory failure in newborn infants. Cochrane Database Syst Rev 2008:CD001340.
6. Hedrick H L. Management of prenatally diagnosed congenital diaphragmatic hernia. Seminars in Fetal & Neonatal Medicine 15 (2010) 21–27
7. Pejovic B, Peco-Antic A, Marinkovic-Eric J: Blood pressure in non-critically ill preterm and full-term 2007;22:249.
8. Bustani P: Congential Diaphragmatic Hernia (CDH)-Neonatal. Neonatal Guidelines. Sheffield Teaching Hospitals – 06.11.18 http://nww.sth.nhs.uk/STHcontDocs/STH_CGP/Neonatology/CongenitalDiaphragmaticHerniaNeonatal.pdf.
9. Embrace guideline for use of inotropes in Neonatal patients (CG1839)