

Postnatal Management of Congenital Diaphragmatic Hernia		Version: 6
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Executive Summary

SUMMARY BOX AND LEVELS OF EVIDENCE¹⁻³¹

After delivery, the infant should be intubated immediately without bag and mask ventilation	D
Aim for preductal saturations between 85-95%	D
Avoid PIP of >25 where possible	D
Consider HFOV if conventional ventilation fails	D
PPHN should be treated with NO	D
ECMO may decrease mortality	D
Surgical repair should be delayed until physiologically stable	D

NATIONAL RECOMMENDATIONS FROM THE CONFIDENTIAL ENQUIRY (MBBRACE)

Recommendations

- Commissioners, service planners and policy makers should consider the establishment of clinical networks that manage the care of babies with CDH. This would include focusing the acute care of these babies on a limited number of centres in order to facilitate the development of:
 - Multidisciplinary teams capable of providing care focussed on both the mother and the baby;
 - A collaborative approach to R&D allowing care to become more evidence based;
 - Sub-specialty expertise in the management of all aspects of care;
 - An agreed 'national information sheet' to provide consistency of information about the condition;
 - Consistent counselling throughout the care pathway;
 - Quality improvement and the sharing of best practice;
 - Collaboration with the proposed national congenital anomalies register to ensure complete case ascertainment;
 - Consensus on the optimal management of the care and treatment for babies diagnosed with CDH and the management of late termination of pregnancy in this group of women.
- Commissioners, service planners and policy makers should consider the development of a service specification for CDH to ensure the service becomes focussed on the needs of the family with, for example:
 - Access to psychological support where applicable;
 - Adequate follow-up arrangements.
 - Organisation of care during the antenatal period in a way that minimises the need for travel;
- There is a continuing need to highlight the importance of documentation to the whole multidisciplinary team involved in the care of women and babies following a diagnosis of CDH.

1. Background

Congenital diaphragmatic hernia (CDH) occurs in approximately 1 in 3000–4000 live births, and is associated with a high overall mortality and a high rate of morbidity amongst survivors. Estimates of the number of cases in the UK vary but it is likely that there are between 200 and 300 new cases annually. Of these up to 70% are likely to be diagnosed antenatally as part of routine screening. Up to a third of all UK cases end in either a spontaneous loss during the pregnancy or an elective termination. A national confidential inquiry on the management of neonates with CDH delivered in 2009-10 identified enormous variation across the UK in the postnatal management of neonates with CDH.[1] The following pathway covers a standard approach based on the recommendations on good clinical practice made by the Topic Expert Group of the committee set for the national confidential enquiry and current available literature. This guideline covers the management of the baby with an antenatal diagnosis of congenital diaphragmatic hernia after delivery. [1-12] This guideline does not cover the antenatal pathway. Further information regarding the antenatal management of CDH and information such as lung head ratios and issues such as FETO are provided in references 3-8.

The key tenets of this guideline are establishing uniformity in practice, and clear pathways for management and escalation in the management of neonates with an antenatal diagnoses of CDH. It must be recognised that even with the best of knowledge and skills human factors play a critical role in management and outcome. For some background please [click here](#) to view the report and recommendations of the National Confidential Enquiry into Congenital Diaphragmatic Hernia conducted by MBBRACE-UK.

An overview is provided in Appendix 1, 2 & 3 to summarise the important tenets of management.

2. Guideline- Preparation prior to delivery

General Principles

A multidisciplinary team should be allocated to attending delivery to include a senior member of staff (usually a consultant). Preparation should be made before hand and if time allows, the care of the baby should be discussed with the parents. Tasks should be allocated to members of the resuscitation team prior to the baby's delivery, identifying responsibilities for airway management, vascular access and medication delivery. Resuscitation equipment should be prepared beforehand by both medical and nursing team members. The labour room resuscitaire should also be thoroughly checked. Below is a list of equipment for the delivery room. Note that surfactant should not be drawn up routinely. This should only be prepared for premature babies who would normally receive this for surfactant deficiency. **Consider discussion with PICU at this stage if the baby is considered to be high risk and possibly requiring ECMO (to facilitate their resource planning).**

List of equipments for delivery room (Including but not limited to Appendix 1A &1B)

- Printed copy of management flow chart
- Pre-cut endotracheal tubes with introducers
- Net-elast hat and ties
- Laryngoscope and blades
- Suction catheter including a Yankuer sucker
- 24G Cannulas (Preferred option for access)
- Single lumen UVC with cord tie and Scalpel (For failure of cannulation or emergency access)
- T- piece flushed through and attached to syringe
- Aliquots of normal saline 10mls/kg (depending on estimated weight of baby)
- Nasogastric tube with purple syringes
- 2 doses of sedation, preferably Fentanyl.
- 2 doses of muscle relaxant, also prescribed according to estimated weight
- 0.9% saline flushes, two 5ml syringes as flush.
- Pulse-oximeter (To be placed on right hand)

Note that resuscitation where possible should be under direction of a consultant or senior personnel experienced in management of such babies.

An estimated weight using the 50th centile for gestation can be used for drug calculation.

Where delivery of a baby with a CDH is suspected preparation of a bed space on the neonatal unit with the appropriate ventilator and delivery of Nitric Oxide must be considered prior to delivery if time allows. If HFOV is needed the Sensormedics oscillator is preferred for full term babies.

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3. Guideline-Delivery room management and stabilisation

Standardised Approach(See figure 1 and Training Video)

Personnel- Ideally resuscitation should be by 2 neonatal resuscitators skilled in neonatal airway and access (one of whom should be a consultant) and an experienced nurse.

Oxygen-Use 100% oxygen for resuscitation and ventilation [12].

Intubate- Intubate immediately without bag and mask ventilation. Ventilation by bag and mask may cause distension of the stomach and must be avoided as it may limit expansion of the hypoplastic lung. Rapid intubation reduces a possible risk of pulmonary hypertension due to prolonged acidosis and hypoxia which may result from delayed intubation. Do not delay intubation whilst obtaining intravenous access [12].

Peak Inspiratory Pressure (PIP) & Positive End Expiratory Pressure (PEEP)

Aim for the minimum PIP that enables adequate chest expansion and ventilation. Ideally this should be ≤ 25 cm of H_2O with a PEEP 5 but clinical judgement is needed here and higher pressures may be needed to produce chest expansion [12].

Monitoring-Attach a saturation probe to the right hand and aim for sats of 80-95% by 15 minutes of age where possible [12].

Decompress the Stomach-An orogastric or nasogastric tube (large bore 8-10) should be placed to decompress the stomach and bowel and left on free drainage. This will help to prevent bowel distension and any further lung compression.

Access-A peripheral venous line should be inserted for sedation and paralysis in delivery suite. In an emergency a UVC may become necessary but where possible umbilical lines should be inserted in a controlled environment i.e. on the neonatal unit.

Sedation and Paralysis-As soon as venous access is established, give a dose of Fentanyl (4microgram/kg) and a paralytic agent. This helps prevent the physiological response to awake intubation which may be uncomfortable raising intracranial pressure, pulmonary pressure, causing bradycardia. Paralysis along with sedation is recommended to ensure the baby is not fighting. . In the absence of IV access, I/M Suxamethonium can be given at a dose of 4milligrams/kg.

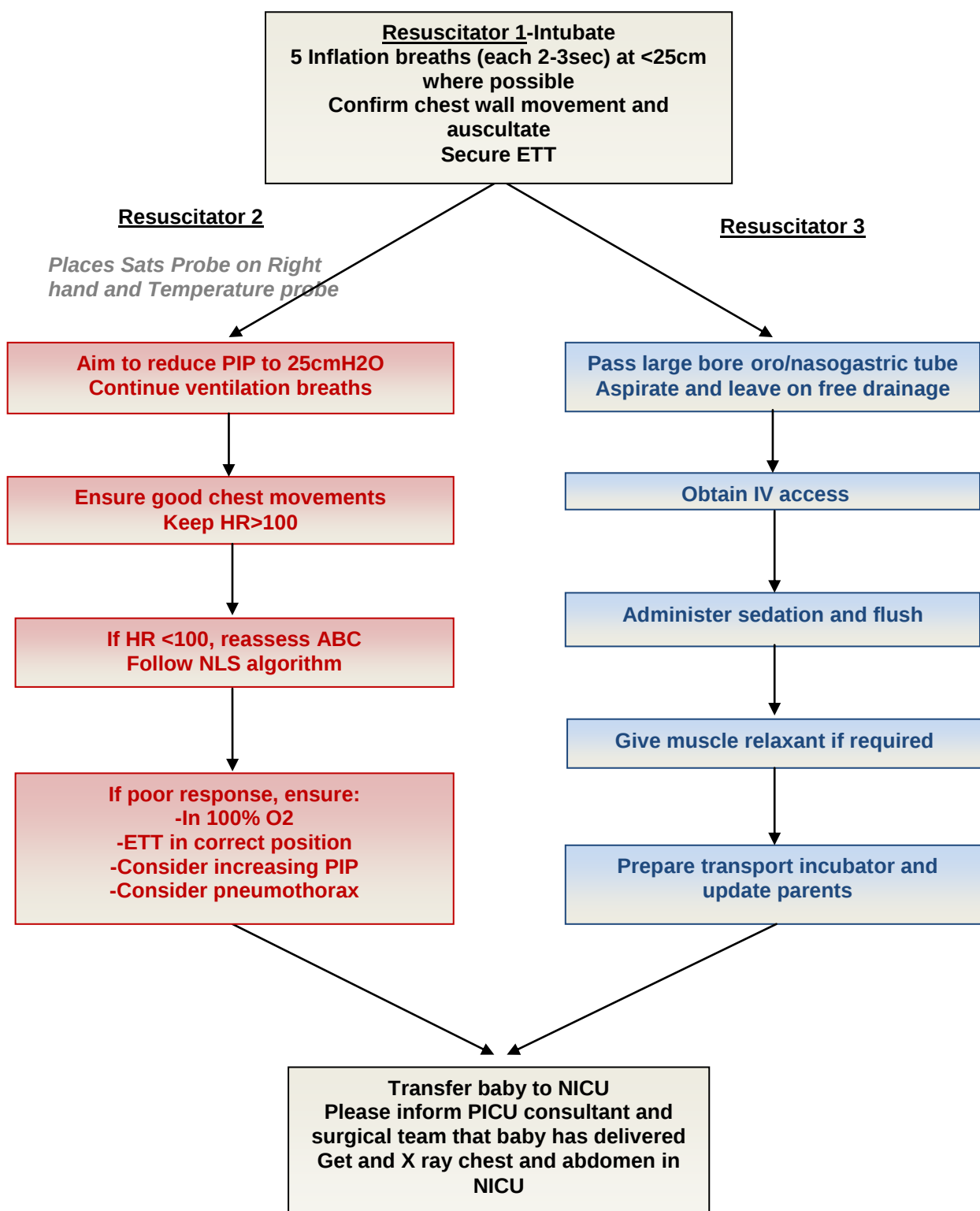
Cardiovascular Assessment- A clinical evaluation of the cardiovascular status is necessary. 10mls/kg of crystalloid can be given 1-2 times at clinical discretion.

Surfactant-Delivery room surfactant for these babies is not recommended.

Temperature- Maintain normothermia [12].

Figure 1

Delivery room management



4. Guideline-NICU Management

Management should be led by the service/on call neonatal consultant.

Ventilation management-General Principles

1) The optimal initial ventilation mode for newborns with CDH varies. There is accumulated evidence that ventilator induced lung injury may have a significant negative impact on outcome in newborns with CDH [13-16]. There is also a risk of precipitating a pneumothorax through use of high pressure ventilation. Having said that persistent hypoxia can result in intractable PPHN. Permissive hypercapnia and gentle ventilation in neonates with CDH has been reported to increase survival [17-20]. Early postnatal ventilator management would be a balance between achieving adequate oxygenation and not driving the lung too hard.

2) Lung Protective Strategy - The European CDH Consortium Consensus recommends adapting treatment to target preductal saturations of **85% and 95%** and a postductal saturation of >70% where possible once of the NNU. This is in keeping with a lung protective strategy outlined in the confidential enquiry into CDH. This will often need high oxygen delivery and even nitric oxide and after stabilisation, FiO₂ should be decreased if preductal saturations are persistently >95% [1, 12].

3) Consideration should be given to the following

Aim for PaCO₂ of between 6-8Kpa (45 and 60mmHg)

Try to limit PIP to 25 cmH₂O or less, and PEEP to 5cm H₂O where possible.

Consider HFOV early in neonates needing higher pressures.

Ensure adequate sedation and paralysis with intravenous morphine (and Midazolam if necessary). Continue paralysis after delivery room management.

Administer a bolus of a muscle relaxant if breathing is asynchronous or ventilation is proving difficult despite the above measures.

A chest X ray should be done as soon as possible after delivery and stabilisation to assess recruitment.

Ensure blood gas analysis occurs regularly and ideally within 1 hour of a significant change to ventilator settings.

Document all changes on the gas chart along with the oxygenation index (OI, OI=mean airway pressure x FiO₂ x 100/PaO₂. [12].

4) PPHN - Anticipate PPHN. Early echocardiography and treatment are very important to guide management here. Indications for nitric oxide are a high oxygen requirement, difference between pre and post ductal saturations and lack of oxygenation on high FIO₂ and inotropes. (See PPHN section)

5) Surfactant - The use of surfactant has not been shown to improve survival. It can be considered at consultant discretion on a case by case basis or where RDS is thought to be an additional pathology as may be the case in a preterm baby with CDH [12].

Ventilation-Which Mode To Use?

There are no data to show the superiority of either conventional or high frequency oscillation as the primary ventilation support. Most babies will start with conventional ventilation [21, 22].

Conventional

It is worthwhile trying conventional ventilation, however if a PIP of over 25cm H₂O is necessary to achieve PCO₂ and saturation levels within the target range despite being in oxygen, consider (HFOV)

High frequency Oscillatory Ventilation (HFOV)

The physiological rationale for use of HFOV derives from its ability to preserve end-expiratory lung volume while avoiding over distension, and therefore lung injury at end-inspiration. HFOV may improve gas exchange, promote uniform lung inflation, reduce barotrauma, and decrease the presence of inflammatory mediators [13, 14, 15, 22].

The indications for HFOV are not clearly defined but it is mostly used as rescue therapy in patients with persisting hypoxemia and hypercapnia on conventional ventilation. [12]

Initial setting; mean airway pressure 2 above conventional MAP setting, frequency 8-10Hz, DP 30-50cmH₂O depending on chest wall vibration to produce visible wobble [12].

The Mean airway pressure should be adjusted to have an adequate expansion of the lungs but avoid over inflation. Over inflation can compromise venous return to the heart and function. This can make PPHN worse.

A chest x-ray should be performed within half an hour to confirm the lungs are not overinflated, as defined by a contra lateral lung expansion such that more than 8 posterior ribs are visible above the diaphragm.

It must be remembered that for big term neonates the Sensormedics may be needed to deliver HFOV. This takes time to set up and if delivery of a baby with a CDH is expected this and the Nitric Oxide should be set up next to the baby.

Hemodynamic management

Assessment and vascular access

Where possible monitor intra-arterial blood pressure using an umbilical artery catheter (UAC) or peripheral arterial line (preferably placed in the right radial arterial line to allow measurement of pre-ductal PaO₂). A central venous line (double lumen UVC) should be inserted in NICU. Aim for a mean arterial pressure (MAP) sufficient to maintain end organ perfusion (MAP = gestational age or MAP = 40 - 45 mmHg in a term baby). Monitor heart rate, capillary refill, urine output and **arterial lactate**.

An echocardiogram should be performed as soon as practicable or within 24 hours to assess ventricular function, pulmonary artery pressure, associated congenital cardiac anomalies and the direction of blood flow across the Ductus Arteriosus (PDA) and Patent Foramen Ovale (PFO)

Stable cardiovascular status is suggested by a combination of the following

- Heart rate within normal range for gestational age
- Central capillary refill $\leq$ 2-3 seconds
- Urine output $\geq$ 1 ml/kg/hr.
- Arterial Lactate $\leq$ 3 mmol/l
- Normal central volume pulse volume

Echocardiography: If symptoms of poor perfusion exist and/or the MAP $\leq$ normal for gestational age request an urgent echocardiogram to differentiate hypovolaemia from ventricular dysfunction and/or PPHN. This will help define the appropriate management strategy. Appropriately experienced and correctly targeted paediatric cardiology advice should be sought.

Management: Consider the cautious administration of 10-20mls/kg of 0.9% saline to augment pre-load (beware previous fluid boluses/hepatomegaly or cardiomegaly as ventricular over-distension can reduce cardiac output and compromise coronary perfusion). A key finding of the confidential enquiry into CDH management was the inappropriate use of fluid boluses without an appropriate objective assessment of intravascular filling status.

If $\geq$ 20ml/kg are required for continued cardiovascular instability consider the use of inotropes, starting with Dopamine (10 mcg/kg/min). If PPHN is thought either clinically or echocardiographically to be a significant contributory factor to circulatory instability start iNO and inotropes/vasopressors (DO NOT delay treatment if there are clinical suspicions of PPHN if an echocardiogram cannot be performed immediately)

Hydrocortisone may be used for treatment of hypotension after failure of conventional treatment and there is inotrope resistant hypotension.

Consider early involvement of PICU if there are signs of significant myocardial dysfunction or severe PPHN as an ECMO referral may be appropriate.

Management of Pulmonary Hypertension

The optimal treatment of CDH associated pulmonary hypertension is one of the major challenges. Support of the systemic circulation and acceptance of variable degrees of right to left shunting (intracardiac) are cornerstones of the therapeutic approach. With severe PPHN it may be necessary to control the direction of ductal blood flow to allow BOTH adequate pulmonary blood flow and optimal systemic oxygen delivery/RV off-loading with a combination of pulmonary vasodilators, inotropes, lusiotropes and systemic vasoconstrictors. This needs to be performed with the assistance of serial echocardiograms and the involvement of a consultant paediatric cardiologist. Serial echocardiograms by an experienced cardiologist may be required. Interpretation of the echocardiogram can be difficult and a consultant cardiology opinion must be sought early if the patient is unstable.

Pulmonary hypertension may be indicated by difficulty in oxygenation, a significant (>10%) difference between Pre and Postductal saturations, or signs of poor systemic perfusion in the face of difficult ventilation. It should also be considered if the Oxygenation Index is over 20. If the ductus arteriosus is closed there will be no pre / post ductal saturation difference, even in the face of severe pulmonary hypertension. Prostaglandin may be required to augment the circulation in this situation if the patient is in cardiogenic shock. Treatment of pulmonary hypertension may require multiple strategies started simultaneously.

1) Optimise PVR and Systemic BP

- a) Volume-10-20mls/kg of saline or HAS can be used but **avoid overfilling** as this can make ventricular function worse.
- b) In PPHN It may well be that a **higher systemic BP** is needed to prevent right to left shunting based on echo assessment to treat PPHN. In addition there may be problems with right ventricular dysfunction.
- c) The **choice of inotropes** is at consultant discretion & may need close liaison with the paediatric cardiology, PICU team. Dopamine or Noradrenaline may be suitable depending on gestation.
- d) Consider use of **Prostaglandin E1** early if there is right ventricular dysfunction with PPHN and a restrictive PDA.
- e) Consider use of **Hydrocortisone** where there is poor response to inotropes.
- f) The use of **Milrinone** in addition to Noradrenaline in right ventricular dysfunction can help offload the right ventricle.

1) Optimise Physiology

- a) **Metabolic Acidosis**-It impairs myocardial function and results in pulmonary vasoconstriction. Treat with sodium bicarbonate or THAM
- b) **Transfusion** can help optimise the oxygen carrying capacity
- c) Optimise **ionised Calcium** and Serum **Magnesium** in support of blood pressure
- d) Maintain axillary temperature between 36.6-37.5C. Avoid **hypothermia** and hyperthermia.
- e) Good sedation and analgesia are important as pain can make pulmonary hypertension worse. **Pain** also causes tachycardia which impairs ventricular filling.
- f) The use of a high **mean airway pressure** or PEEP can impair venous return to the right heart making PPHN worse.
- g) Review the chest Xray to look at the heart size and lung expansion.

Nitric Oxide & Vasodilators

1. Consider the early use of iNO (10-20 ppm) when PPHN is suspected or confirmed (**Hypoxia with a pre-post ductal saturation difference of $\geq 10\%$ or echocardiographic evidence of significantly raised (near/supra-systemic) pulmonary artery pressures**). The efficacy of iNO should be assessed ≤ 1 hour (a $\geq 20\%$ reduction in either PA pressures or OI). Although a randomised controlled trial did not demonstrate a beneficial effect of iNO on mortality [23], an immediate short term improvement in oxygenation may be beneficial as a bridge to ECMO.
2. Consider IV prostaglandin E1 in cases of suprasystemic pulmonary arterial pressure and right to left shunting through the foramen ovale on echo especially if there is right ventricular dysfunction and a restrictive PDA.
3. IV Magnesium is a pulmonary vasodilator and can be administered in cases of refractory pulmonary hypertension. It can make things worse by causing systemic hypotension through vasodilatation and may need escalation of vasopressors. It's use should be discussed with the consultant and closely monitored.
4. Sildenafil and other pulmonary vasodilators (Prostacyclin) for PPHN have been used on a case by case basis to treat PPHN. There is no evidence that these help in the acute phase in infants with CDH however they may benefit a baby with suprasystemic pulmonary hypertension and after consultation with a cardiologist use on a case by case basis may be warranted. [12]

Sedation and Analgesia

Steps (12)

Keep the environment quiet! Avoid handling where possible. Preoxygenate prior to handling by increasing FiO_2 to 1.

Morphine given as a continuous infusion is the sedation of choice on NICU (additional Morphine boluses may be required). Sedation should be titrated using clinical assessment (heart rate and blood pressure). Be aware that this might cause hypotension and the need for increased inotropic support.

Midazolam may be an appropriate additional sedative agent where asynchrony with ventilation and agitation causing hypoxaemia are encountered.

Rectal Chloral hydrate is also an option when NBM or orally when fed.

The use of paralysis is almost universal in neonates with PPHN associated with CDH however its use has side effects. Consider using and weaning infusions to the lightest dose necessary to aid sedation and stopping if not needed.

Extracorporeal Membrane Oxygenation (ECMO)

ECMO has been reported to improve the survival of infants with CDH [24, 25]. The utilisation of ECMO ranges from 15 to 40% [26]. The overall survival of CDH following ECMO is 51% in this population [27]. In many centres, ECMO is considered in infants with CDH if there is evidence of an adequate amount of lung parenchyma suggested by a period of adequate preductal oxygenation and/or ventilation. The national confidential enquiry recommends that in cases where the baby could be adequately stabilised and the degree of pulmonary hypoplasia was felt to be compatible with long term survival ECMO should have been considered. The case should be discussed early with an ECMO Centre. Following discussion with an ECMO Centre, the case for/against ECMO should have been discussed with parents. The reasons for its use, its limitations and why it was/was not advisable for the treatment of their baby should have been documented. It must however be remembered that wider issues might preclude ECMO. Other co morbid conditions need to be considered before referral for ECMO. These include prematurity, presence of significant Intraventricular haemorrhage, lethal malformations and congenital anomalies and syndromic diagnosis with poor prognosis. ECMO is only worth considering if the baby has lungs which are compatible with survival. In babies with severe pulmonary hypoplasia or where there are other congenital abnormalities ECMO does not play a useful role. If the baby has never had a preductal PaO₂ above 4-5kPa and ventilation has been difficult from the beginning despite maximal treatment then ECMO may be inappropriate. Please note that despite use of ECMO, mortality remains high. ***Babies are to be discussed for ECMO alongside careful multidisciplinary discussion with the PICU and surgical consultants.*** It is essential that if a patient is not responding to treatment, or deteriorating, then ECMO is considered early and the patient is discussed with the UHS PICU consultant in a timely manner, as they may be in a position to offer help with stabilisation, or help facilitate referral and transfer for ECMO.

ECMO options include:

- Early transfer on conventional ventilation by Lead ECMO centre for subsequent consideration of ECMO in stable child with a high/worsening OI
- Transfer to SGH PICU for consideration of initiation of ECMO prior to retrieval by Lead ECMO centre
- Stabilisation of and cannulation of child on NICU by ECMO transport team from Lead Centre

Criteria for ECMO referral [12]

Indicators that should prompt a referral and discussion are as below (not limited to these criteria individually)-

- $OI \geq 40$ or inability to maintain preductal saturation of $>85\%$ or postductal of $>70\%$
- $OI >20$ consistently, with signs of cardiogenic shock requiring high dose inotropic support.
- Increased $PaCO_2$ with $pH < 7.15$ despite optimising ventilator management
- Systemic hypotension resistant to fluid and inotropic therapy with urine output of $<0.5\text{ml/kg/hr.}$ for at least 12 to 24 hours
- Inadequate oxygen delivery with metabolic acidosis as measured by lactate $>/- 5$ and $pH < 7.15$
- Rapid deterioration or severe ventricular dysfunction [28]
- Severe air leaks unresponsive to other therapies [28]
- No contraindication to ECMO (see above)

Fluid management and Parenteral Nutrition

- Fluid restriction ($40\text{-}60\text{mls/kg/day}$) should be considered in the first 24 hours. Strict input and output chart should be kept.
- The use of paralysis is likely to make these neonates oedematous.
- Total parenteral nutrition (TPN) should be started within 24 hours once suitable central access has been obtained. These babies are not likely to be fed until after surgery.
- Monitor electrolytes daily to start with. If stable, they can be checked with TPN bag changes.
- Consider diuretics where appropriate in case of a positive fluid balance. Aim for diuresis of $1\text{-}2\text{ml/kg/hr.}$ A low albumin may necessitate the need of 20% Albumin on a case by case basis. It is important to monitor renal function closely in such situations.

Timing of Surgery

Consensus exists that repair of the diaphragmatic defect should be done as a semi-elective procedure once the baby has stabilised. Physiological stabilisation [11] has been defined as

- Mean arterial blood pressure normal for gestational age.
- Preductal saturation level of 85-95% in FiO₂ <50%.
- Arterial lactate <3mmol/L.
- Urine output >2ml/kg/hr.

A key area is making a judgement that pulmonary pressures are stable and that there is good myocardial function. The decision regarding operation should be a joint decision between the neonatal surgical consultant and the consultant neonatologist.

An anaesthetic review should be encouraged prior to surgery.

Echocardiography is helpful in allaying any concerns in this regard if there is any uncertainty. It is also important to ensure that we are prepared to manage any deterioration during surgery. Keeping Nitric Oxide attached and ready to run as well as inotropes ready is best done prior to surgery. Surgery can be performed on HFOV if the above criteria are met.

The following should be ensured prior to surgery

1. Preanaesthetic review by Paediatric anaesthetic consultant
2. Preoperative echo if concerns regarding pulmonary hypertension
3. Consent and discussion with parents
4. FBC and clotting done and results checked
5. Cross-match of 2 units of blood
6. Appropriate central and peripheral access (checked and working)
7. Functioning arterial access
8. Nitric oxide available and run though ready to run
9. There is no evidence of evolving sepsis

5. Guideline-Post operative management

- Standard post operative care to include blood tests (FBC and U&E).
- Obtain a chest X-ray. This helps assess the air pocket and mediastinum.
- In the immediate postoperative period, there will be air in the pleural space but this is not usually under tension. There will usually still be some mediastinal shift away from the lesion but this may be less than pre-op. This pleural air can be mistaken for a tension pneumothorax but it rarely needs draining and will be replaced by fluid or the lung will expand into the space over a few days. Needle thoracocentesis or chest drainage MUST NOT be performed without consultation between a senior surgeon and neonatologist. The instability of the mediastinum can impact upon blood pressure and postoperative PPHN can be challenging to manage. Careful management of blood pressure & echocardiography to evaluate filling and cardiac function is important at this stage.
- Optimising blood pressure, use of Nitric Oxide (even for initial non responders) sedation and analgesia are key in managing postoperative PPHN.
- These infants may have an increasing ventilator requirement in the immediate postoperative period. Be prepared to escalate ventilation. The same applies for inotropic support. Monitor blood pressure and start inotropes sooner rather than later.
- Weaning off Nitric Oxide can be challenging and some neonates may need to start Sildenafil to allow transition of it. In some cases persistently elevated pulmonary artery pressures may need to be managed using oral Sildenafil after the neonate has been extubated.
- Need for chest drain or thoracocentesis later on is rare unless there is a large pleural effusion compromising lung function and ventilation. This should be discussed with surgical team where possible.
- Perioperative antibiotics should be as per protocol.
- Enteral feeding should be started as guided by the surgical team. The use of antireflux medication should be guided by the surgeons as Reflux is a common problem.
- Ensure stress for the family is minimized: Parents should be kept informed of baby's condition and progress. They should be given the opportunity to speak with the surgeon/doctors/nurses.

6. Prognosis and Follow Up

The Confidential inquiry into management of CDH has advocated for a family centred approach. The reported survival rates for CDH range from 30 to 80% depending on case selection [29]. This averages out to about 50% in population based studies [30]. The prognosis is significantly worse when other associated congenital anomalies or genetic abnormalities are present. [31] The prognosis for right sided diaphragmatic hernias is poorer as compared to left sided cases. [31,32] **Discussions with the parents about prognosis and of management, and care plans must be carefully documented in the patient record.** Chronic lung disease, ongoing pulmonary hypertension, Neurodevelopmental problems, sensorineural hearing loss and gastroesophageal reflux are common problems in survivors. The need for respiratory support in the NICU and home necessitate involvement of the Paediatric Respiratory team. Ongoing pulmonary hypertension may need involvement of the Paediatric Cardiology team.

Follow up of CDH patients would be organised with the surgical and neonatal team. Depending on the individual needs of each patient paediatric respiratory, cardiology and Neurodevelopmental follow up may also be needed.

For patients discharged to their local units follow up would be with their local paediatricians and the surgical team. Multidisciplinary input from specialist services might be needed locally.

7. Process for Monitoring Compliance/Effectiveness

Key aspects of the procedural document that will be monitored:

What aspects of compliance with the document will be monitored	What will be reviewed to evidence this	How and how often will this be done	Detail sample size (if applicable)	Who will co-ordinate and report findings (1)	Which group or report will receive findings
CDH Management Pathway	Audit of delivery room management and postnatal care	3 years		Consultant or Neonatologist Consultant Paediatric Surgeon	Neonatal and Paediatric Surgical consultants

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Appendix 1A: Equipment for Airway & Access in Delivery Suite

- Pre-cut endotracheal tubes with introducers
- Net-elast hat and ties
- Laryngoscope and blades
- Suction catheter including a Yankuer sucker
- 24G Cannulas
- Single lumen UVC with cord tie and blade (For failure of cannulation)
- T- piece flushed through and attached to syringe
- Aliquots of normal saline 10mls/kg (depending on estimated weight of baby)
- Nasogastric tube with purple syringes

Appendix 1B: Drugs for Sedation and Paralysis

- Medications
- 2 doses of sedation, preferably Fentanyl. (4 microgram/kg)
- 2 doses of muscle relaxant, also prescribed according to estimated weight
- 0.9% saline flushes, two 5ml syringes as flush
- If no IV access is obtainable and paralysis is required Suxamethonium can be given at a dose of 4mg/kg

Appendix 2- Imminent Delivery of a baby with CDH (Preparation s key!!!) Human Factors



Communication

Prebrief

Who is team leader?

Delegate who is doing ABC, checking the equipment and the resuscitaire

Is a consultant on the way if not already there?



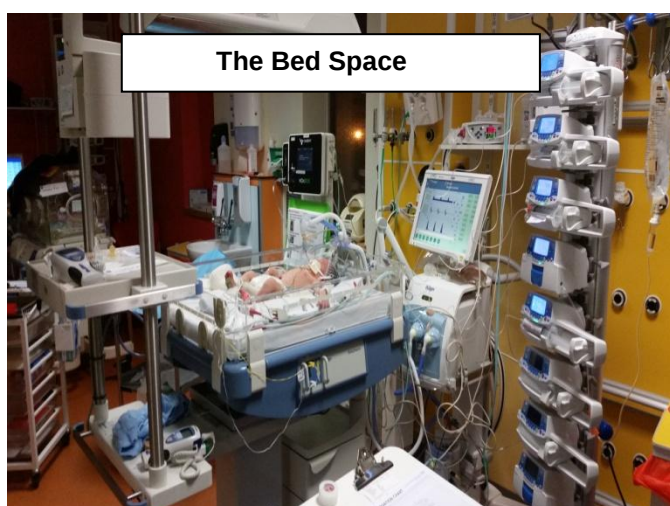
The Resuscitaire

Have you set PIP and PEEP?

Does it have oxygen/enough oxygen?

Does your laryngoscope work?

Do you have capnography?



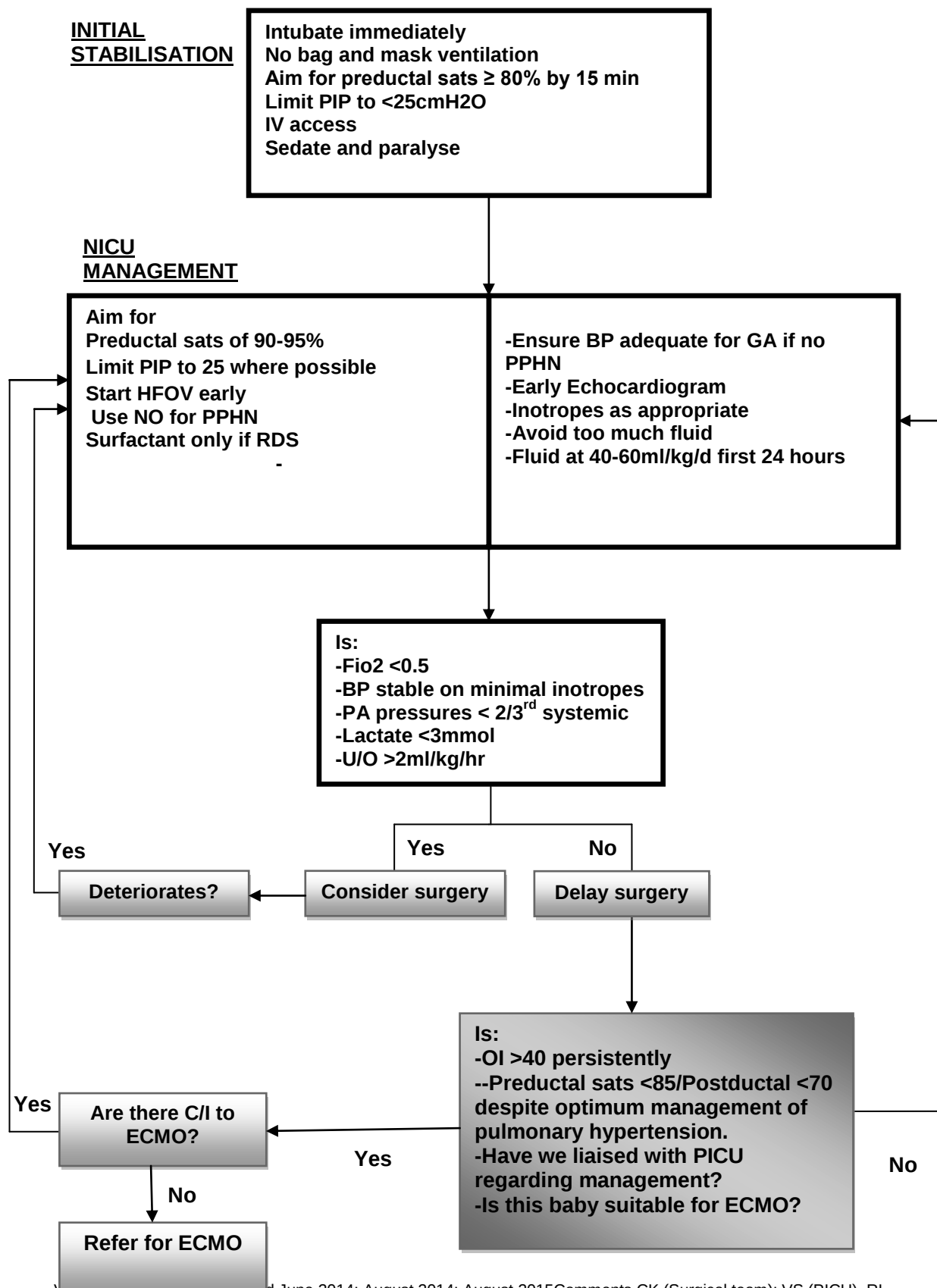
The Bed Space

Is a bed space with the appropriate ventilator and equipment ready?

Has the Nitric Oxide checked and run through?

Is a Sensormedics available?

Is the PICU consultant/ PICU aware?

Appendix 3: CDH Management Pathway

Monitoring Information

Postnatal Management of Congenital Diaphragmatic Hernia

Version: 6

The Trust strives to ensure equality of opportunity for all, both as a major employer and as a provider of health care. This document has therefore been equality impact assessed to ensure fairness and consistency for all those covered by it, regardless of their individual differences, and the results are available on request.

Approval Committee:	Neonatal Governance
Date of Approval:	Insert Date
Ratification Committee:	Insert Name of Committee (Policy Ratification and Monitoring Group (PRAMG) for Level 1 documents)
Date of Ratification:	Insert Date
Signature of ratifying Committee Group/Chair:	Insert Signature or name (Chair of PRAMG if Level 1 document)
Lead Name and Job Title of originator/author or responsible committee/individual:	Insert name and job title of responsible individual/author or responsible committee
Policy Monitoring (Section 6) Completion Date:	Insert Date
Policy Monitoring to be presented to responsible committee or PRAMG:	Insert Date or Month applicable
Target audience:	Neonatal, PICU, Maternity, Paediatric Surgery
Key words:	
Main areas affected:	Trust wide for Level 1 documents
Summary of most recent changes if applicable:	None
Consultation:	As stated in the footer
Equality Impact Assessment completion date:	
Number of pages:	25
Type of document:	Guideline
Does this document replace or revise an existing document	No
Should this document be made available on the public website?	No
Is this document to be published in any other format?	No