

Infants with Congenital Diaphragmatic Hernia

Pre-Operative Management Guidelines

*****This document is a general guideline and does not represent a professional care standard governing providers' obligation to patients. Care is revised to meet individual patient needs. This is a quality improvement document and should not be part of a patient's medical record.***

OBSTETRIC MANAGEMENT

1. Delivery at UNC
2. Prenatal consult with Neonatology & Pediatric Surgery
3. Fetal echocardiography as determined by Maternal Fetal Medicine
4. Cord blood for type & cross
5. Cord blood for karyotype & genetic testing

DELIVERY ROOM MANAGEMENT

1. If respiratory support needed endotracheal intubation. (Goal is to avoid PPV by face mask or Neo puff)
2. Place oxygen saturation monitor on right arm (preductal saturation)
3. If intubated:
 - a. Begin resuscitation with FiO₂ 1.0 and wean to keep preductal SpO₂ >80%.
 - b. Ventilate per NRP guidelines with every effort to avoid PIPs >25.
 - c. Initiate ventilation with PEEP 5 cmH₂O.
 - d. Place replogle tube (at least 10 Fr) to low continuous suction.
 - e. Notify Pediatric Surgery, PICU, ECMO on child's arrival to NCCC.
 - f. Consider surfactant in infants < 34 weeks gestation.

ADMISSION MANAGEMENT AFTER DELIVERY ROOM (NICU or PICU)

1. Mechanical Ventilation Strategy
 - a. Equipment
 - i. Drager/Servo-U Ventilator
 - ii. Cerebral & Renal NIRS (PICU)
 - iii. Capnography/Transcutaneous CO₂ monitoring
 - iv. Inhaled nitric oxide
 - v. HFOV: 3100 A + oxygen analyzer
 - b. Clinical Goals
 - i. Goals may change based on provider discussion
 - ii. pH 7.25-7.4
 - iii. PaCO₂ 40-60

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- iv. $\text{PaO}_2 > 80$
- v. Pre-ductal $\text{SpO}_2 > 92\%$
- vi. Oxygenation index < 20
 - 1. $\text{OI} > 20$, consider iNO (see iNO section below)
 - 2. $\text{OI} > 30$, consider starting HFOV
 - 3. $\text{OI} > 40$, consider ECMO
- c. Manual (bag or Neopuff) Positive Pressure Ventilation
 - i. Avoid if possible, may need to use if $\text{SpO}_2 < 80\%$ or unresponsive to ventilator changes
 - ii. If manual PPV is necessary, use $\text{PIP} < 28$
 - iii. Requires a verbal order from provider.
- d. Conventional ventilation with Drager/Servo-U
 - i. Mode: AC-VG (Drager) or PRVC (Servo)
 - ii. Initial settings
 - 1. Tidal volume 4-5 ml/kg
 - 2. PEEP 5
 - 3. IMV rate: 40
 - 4. I-time 0.3-0.45 sec
 - 5. FiO_2 : 100%
 - 6. Max PIP: 28 mmHg
 - iii. See Appendix for titration/management of CMV settings
- e. HFOV
 - i. Escalate ventilator to HFOV if:
 - 1. Patient requiring $\text{PIP} \geq 28$ during PPV or mechanical ventilation
 - 2. Persistent pre-ductal $\text{SpO}_2 < 92\%$
 - 3. $\text{PaCO}_2 > 60$ on optimal conventional ventilation with adequate blood pressure support and sedation
 - 4. Oxygenation index (OI) > 30
 - ii. Initial settings (for ≥ 35 weeks gestation)
 - 1. Hz 10
 - 2. MAP 16-22 (using conventional vent MAP, add 1-2 above)
 - 3. Amp 30-60 (based on physical exam, chest “wobble”)
 - 4. Bias flow 20
 - iii. See Appendix for HFOV management strategies, monitoring, and suctioning guidelines
- 2. Place pre-and post-ductal saturation monitors (goal pre-ductal $> 90\%$)
- 3. Place End tidal CO_2 monitor (Transcutaneous CO_2 monitor if on HFOV) (goal 45-65).
- 4. Total fluids begin at 80 ml/kg/day, D10W with calcium
- 5. Initial ABG within 15 minutes of NCCC admission (see labs)

6. Correlate ABG with CO₂ monitor
7. After weighing, securing monitoring devices and nurse assessment, immediately place UAC and double lumen UVC
8. Lines attempted and initial CXR within 1 hour of admission.
9. ECHO to assess structure, function, degree of pulmonary hypertension and line placement (after lines are placed)
10. For patients arriving from outside hospitals, consider direct admission to PICU if patient already on maximum support.
 - a. PIP >28 on conventional ventilation
 - b. PEEP \geq 8 on 100% FiO₂
 - c. MAP >20
 - d. Oxygenation Index (OI) > 30
 - e. Most recent ABG with pH < 7.20, PaCO₂ >50, or PaO₂ < 80
 - f. Can consult NCCC for line placement if patient without central access.

NCCC/PICU MANAGEMENT

1. Team Management
 - a. Infants with CDH require close coordination between the NCCC, Pediatric Surgery, and PICU. The team should be notified on admission and with any alterations in status requiring consideration of a change in ventilator mode, type of ventilator, initiation of inotropes, neuromuscular blockade, introduction of iNO or ECMO discussion.
 - b. Daily team meeting to include NCCC, Pediatric Surgery, PICU (as indicated), bedside nurse and respiratory therapist.
2. Sedation
 - a. Fentanyl 2-4 mcg/kg/hr or Morphine 0.025 – 0.05 mg/kg/hr
 - b. Maintain minimum stimulation.
 - i. Minimal stimulation nursing care
 - ii. Bolus sedation prior to procedures, suctioning, or touch
 - iii. Ear shields
 - c. Avoid neuromuscular blockade if possible. However, should initiate neuromuscular blockade if the patient oxygenation/ventilation/hemodynamic goals are refractory to respiratory and hemodynamic support. Also consider neuromuscular blockade if there are frequent desaturations.
 - i. If neuromuscular blockade is started, add lacrilube for eyes.
 - ii. Place foley
 - iii. Increase ventilator rate if necessary
3. Circulatory Support
 - a. Maintain mean UAC blood pressure at 40-55 mmHg.
 - b. For hypotension, give a fluid bolus of NS 10 ml/kg. May repeat once. If unresponsive to fluid, start inotropic support.

- c. Inotropes.
 - i. Epinephrine at 0.05 mcg/kg/min (max dose 1.0 mcg/kg/min)
 - ii. Milrinone 0.5 mcg/kg/min
 - iii. May require both medications
- d. If BP unresponsive after initiation of fluid and pressors consider stress dose hydrocortisone.
- e. Maintain ionized calcium within normal range (4.5-5 mg/dL). Use calcium gluconate boluses 100 mg/kg as needed. (can give calcium chloride 20 mg/kg if in PICU)
- f. Consider NaHCO₃ administration if there is adequate ventilation and blood pressure support, but there is persistent acidosis or difficulty oxygenating with significant pre- and post-ductal saturation difference.
 - i. Bolus 1-2 meq/kg NaHCO₃ given over 20 min (or per unit protocol)
 - ii. Extreme caution with bicarbonate as it can lead to increased intracellular CO₂ and acidosis, especially in infants with inadequate ventilation.

4. Labs

- a. CBC with diff, blood culture, type & screen, glucose, ABG with ionized calcium and lactate within 15 min of admission
- b. ABG q2 at a minimum. Add hemoglobin, ionized calcium, and lactate to the ABG q6
- c. CBC with diff daily
- d. Pro-BNP/BNP should be measured daily until pulmonary hypertension has resolved
- e. Chem 10 daily (Na, K, Cl, CO₂, BUN, creatinine, glucose, Mag, Phos)
- f. Target ranges for Blood gases: pH >7.25, PaCO₂ 40-65, PaO₂ 60-100

5. Ventilator Management

- a. Clinical goals:
 - i. pH 7.25-7.40
 - ii. PaCO₂ 40-60
 - iii. PaO₂ >80 preductal, >60 post ductal
 - iv. Pre-ductal SpO₂ >92%
- b. See Appendix for Mechanical Ventilation titration guidelines for conventional ventilator.
- c. Consider escalation to HFOV if requiring PIP ≥ 28, Persistent pre-ductal SpO₂ < 92%, PaCO₂ > 60, or OI >30
 - i. Initial settings (for ≥ 35 weeks gestation)
 - i. Hz 10
 - ii. MAP 16-22 (using conventional vent MAP, add 1-2 above)
 - iii. Amp 30-60
 - iv. Bias flow 20
 - ii. See Appendix for HFOV management strategies, monitoring, and suctioning guidelines.
- d. Transitioning back to conventional ventilation from HFOV (prior to CDH repair)
 - i. Physiological stabilization for at least 12 hours
 - ii. Written order from attending/fellow physician

- iii. Minimum HFOV settings: Amp <35, MAP <16, Hz \geq 10
- iv. pH 7.28 – 7.4, pCO₂ <60, FiO₂ <60%

6. Inhaled Nitric Oxide

- a. *Patient must have had an echocardiogram prior to initiation of iNO*
- b. Consider iNO if echocardiographic evidence of PPHN, Oxygenation Index > 20 (see notes below), or pre-ductal saturation <85%.
- c. Do not start iNO if patient has evidence of isolated left ventricular dysfunction.
- d. Start iNO at 20 ppm per iNO protocol.
- e. Check methemoglobin levels and consider weaning per iNO protocol.
- f. Once on iNO check ECHO once within 12 hours of initiation. Then check ECHO as needed based upon clinical status of the patient
- g. Weaning iNO
 - i. Initiation of iNO wean requires provider order.
 - ii. Wean from 20 ppm by 5 ppm every 6 hours until 5 ppm is reached. Then wean iNO by 1 ppm every 6 hours until off.
 - iii. Consider starting sildenafil when transitioning off iNO.

7. Echocardiograms

- a. Should be obtained shortly after birth and DOL 1 (the day following admission)
- b. Repeat as needed based on changes in hemodynamic status, refractory hypoxia, or after the addition of a vasopressor or pulmonary vasodilator.
- c. The ECHO is a valuable tool in managing PPHN related to CDH. ECHO can assess the severity of pulmonary hypertension, estimate RV pressure, and determine the degree and direction of shunt at the ductus arteriosus and the atrial level. The ECHO can also assess the function of the LV and RV in the face of severe pulmonary hypertension or less commonly LV dysfunction.

8. Studies

- a. While on CV daily CXR to assess for inflation, pneumothorax, ETT placement.
- b. On HFOV at least daily but consider twice daily to assess for inflation, pneumothorax.
- c. Baseline HUS if escalation beyond conventional ventilation (in preparation for possible ECMO consideration)

9. Nutrition

- a. Initial nutrition should be delivered by TPN. The TPN should not deliver more than 80 kcal/kg/day until respiratory/ventilatory parameters and pulmonary vascular pressures are stable. Start on DOL 2. On ECMO, may reduce to 60 kcal/kg/day. Maximize acetate in TPN for metabolic acidosis.
- b. Consider sodium acetate as arterial line fluid based on metabolic status.

10. Genetics

- a. Cord blood should be sent for karyotype and microarray.
- b. Consult Genetics early if a major cardiac defect or physical findings are present to suggest the presence of an underlying genetic abnormality or syndrome. CDH is common in the context of many syndromic and other genetic disorders.

ECMO

1. Alert ECMO team (NCCC, Pediatric Surgery, PICU) for evaluation if patient has failed conventional ventilation and proceeded to HFOV, iNO, or both.
2. Once HFOV or iNO are initiated, order a head ultrasound to rule out intracranial bleed.
3. General ECMO inclusion criteria
 - a. Birth weight ≥ 2 kg
 - b. No major cardiac anomalies or bleeding diathesis
 - c. No IVH > Grade II
 - d. $OI \geq 40$ on 2 consecutive blood gases an hour apart.
4. General guidelines for management while on ECMO, pre-repair
 - a. pH 7.35-7.45
 - b. $PaO_2 \geq 60$
 - c. PCO_2 40-45
 - d. Discontinue iNO
5. ECMO ventilator management
 - a. Servo U – SIMV/PCPS, PC 10 (total PIP 15-20), rate 10, PEEP 5-10, PS 10, iTime 1 sec, FiO_2 40%
 - b. Emergency vent settings posted on ventilator.
 - c. “Tubing compliance off” sign posted if applicable.
 - d. Lung conditioning
 - i. Hold 24 hours post-repair
 - ii. Scheduled Q6 (can increase frequency with change in order from provider)
 - iii. Consider IPV for secretion management if needed.
 - iv. No lung conditioning of patient is on PIP >20, RR 25, or per provider order.
 - e. End tidal CO_2 monitoring not indicated until ECMO trial-off.

APPENDIX

The following are general guidelines for ventilator titration and management for CDH patients. Clinical judgement may require changes for individual patients.

NOTES

Oxygenation index (OI) = (MAP x FiO₂ x 100) / PaO₂

Conventional Ventilation:

Settings/Ranges for CDH Conventional Ventilation Pathway

Mode	Tidal Volume	PEEP	IMV Rate	I-time	FiO ₂	Pmax
Drager: AC-VG Servo: PRVC	4-5 cc/kg	5	35-45	0.3-0.45	100%	28

Conventional Ventilator Management Pathway

Oxygenation	PEEP	FiO ₂
SpO ₂ < 92%	If FiO ₂ is 100%, increase PEEP by 1 if chest x-ray reveals under inflation	Increase FiO ₂ by 5% until reached 100%
SpO ₂ ≥ 92%	FiO ₂ < 60%, decrease PEEP by 1 if clinically indicated on chest x-ray. *Minimum PEEP 5	Decrease FiO ₂ by 5% q1h to maintain goal pre ductal saturations.

**All ventilator changes should only be made in discussion with the provider.*

HFOV

Settings/Ranges for CDH HFOV Pathway

Age	Hertz	Map	Amp	Bias Flow
≥35 weeks GA	10	16 (min) - 22(max) ***Utilize conventional MAP + 1 or 2 above	30(min) - 60(max)	20

HFOV Management Pathway

Oxygenation	Mean Airway Pressure (MAP)	FiO2
SpO ₂ < 92%	If FiO ₂ is 100%, increase MAP by 1-2 every 30 minutes until max range is achieved. May need to increase based on discretion of provider & to maintain SpO ₂ > 92% *Change in chest x-ray	Increase FiO ₂ by 5% until reached 100%
SpO ₂ ≥ 92%	FiO ₂ < 60%, decrease MAP by 1-2 with scheduled ABG until minimum range is achieved. Decrease based on ABG frequency or provider's order. *Change in chest x-ray	Decrease FiO ₂ by 5% until reached 60% Decrease based on ABG frequency or provider's order

****All ventilator changes should only be made in discussion with the provider.***

****Increase/Decrease MAP 1-2cm when chest x-ray shows under expansion (less than 8 ribs visible) or over-inflation (>9 ribs visible)***

HFOV Patient Assessment/Monitoring

- Chest Wiggle: visible bilateral vibration noted from the nipple line to the umbilicus.
- Auscultation: denote changes in the intensity of the piston sounds. Breath sounds cannot be heard.
- ABGs: minimum every 2 hours until clinical goal ranges achieved.
- Vitals
- Transcutaneous CO₂ Monitoring

Arterial Blood Gas (ABGS)

- Obtain an initial ABG prior to placing patient on HFOV & 1hour after HFOV initiation.
- Obtain an ABG 1 hour after any setting adjustments are made to HFOV.
- Obtain ABGS q2 or per MD order.

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Chest Radiograph (Chest Xray)

- Baseline chest x-ray if any escalation beyond CMV
 - a. HFOV (aim for 8 ribs expanded)
- Daily chest x-rays to assess for PTX, ETT placement until repair.
- Obtain a chest x-ray with any acute changes in patient's condition.

Suctioning HFOV

- Lavage suction prior to placing patient on HFOV.
- Suction will be done once a shift with an in-line suction catheter once HFOV initiated.
- Suction is indicated for diminished chest wall movement (no wiggle), elevated pCO₂, worsening oxygenation (may indicate ETT obstruction, or visible secretions in the airway).

Possible adverse effects

- Hyperinflation
- Decreased cardiac output:
 - a. Indications:
 - Tachycardia
 - Decreased peripheral pulse.
 - Decrease in blood pressure.
- Pneumothorax:
 - a. Indications:
 - Change in ABG
 - Decrease in FiO₂ saturations.
 - Decreased chest wiggle on affected side.