Newborn Critical Care Center (NCCC) Clinical Guidelines

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 should be revised to meet individual patient needs.

OBSTETRIC MANAGEMENT

1. Delivery at UNC
2. Prenatal consult with Neonatology & Pediatric Surgery
3. Fetal echocardiography as determined by Maternal Fetal Medicine
4. Obtain cord blood for ABO/Rh
5. Obtain cord blood for karyotype/microarray and DNA extract and hold.

DELIVERY ROOM MANAGEMENT

1. If infant requires respiratory support, intubate immediately - goal is to avoid PPV (face mask or Neo puff
2. Place oxygen saturation monitor on right arm (preductal saturation)
3. If intubated:
    a. Begin resuscitation with FiO_2 1.0 and wean to keep preductal SpO_2 > 80%
    b. Ventilate per NRP guidelines with every effort to avoid PIPs > 25 mmHg
    c. Initiate ventilation with PEEP 5 cm H_2O
    d. Place replogle tube (at least 10 Fr) to low continuous suction
    e. Notify Pediatric Surgery, PICU and ECMO team upon infant'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 *(if infant is in PICU)*
    iii. Capnography/Transcutaneous CO_2 monitoring
    iv. Inhaled nitric oxide
    v. HFOV: 3100 A + oxygen analyzer

b. Clinical Goals *(may change based on provider discussion)*
    i. pH 7.25 - 7.4
    ii. $PaCO_2$ 40-60 mmHg
    iii. PaO_2 > 80 mmHg
    iv. Pre-ductal SpO_2 > 92%
    v. Oxygenation index < 20
        1. OI > 20, consider iNO (see iNO section below)
        2. OI >30, consider starting HFOV

3. OI >40, consider ECMO

c. Manual (bag or Neopuff) Positive Pressure Ventilation
    i. Avoid if possible, may need to use if SpO_2 < 80% or unresponsive to ventilator changes
    ii. If manual PPV is necessary, use PIP < 28 mmHg
    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 cm H_2O
        3. IMV rate: 40
        4. I-time 0.3 – 0.45 sec
        5. FiO_2: 1.0
        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 PIP ≥ 28 mmHg during PPV or mechanical ventilation
        2. Persistent pre-ductal SpO_2 < 92%
        3. $PaCO_2$ > 60 mmHg 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 “wiggle”)
        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) with goal 45 – 65 mmHg
4. Initial ABG ideally within 15 minutes of NCCC admission (see Labs)
5. Correlate ABG with CO_2 monitor
6. After weighing, securing monitoring devices and nurse assessment, immediately place UAC and double lumen UVC
7. IV fluids – begin with total fluids at 80 mL/kg/day infusing D10W with calcium gluconate (200-400 mg/kg/d)
8. Attempt to place umbilical lines and obtain 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 mmHg on conventional ventilation
    b. PEEP $\geq$ 8 cm H_2O on FiO_2 1.0
    c. MAP > 20
    d. Oxygenation Index (OI) > 30
    e. Most recent ABG with pH < 7.20, $PaCO_2$ > 50 mmHg, or PaO_2 < 80 mmHg
    f. Consider consulting NCCC for line placement if patient does not have 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, neuromuscular blockade should be initiated if the patient's 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 lubricant for eyes
        ii. Place foley catheter
        iii. Increase ventilator rate if necessary
3. ***Circulatory Support***
    a. Maintain mean arterial blood pressure of 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:
        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
    c. If BP unresponsive after initiation of fluid and inotropes consider stress dose hydrocortisone.
    d. 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.)

e. Consider $NaHCO_3$ 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 $NaHCO_3$ given over 20 min (or per unit protocol)
    ii. Extreme caution with bicarbonate as it can lead to increased intracellular CO_2 and acidosis, especially in infants with inadequate ventilation

4. ***Labs***

LABORATORY EVALUATION SCHEDULE	
ADMISSION (ideally within 15 minutes)	ABG with iCa, lactate CBC with differential Type & Screen *(and ABO/Rh if not sent from cord blood)* Blood culture Glucose
DAILY	CBC with differential Chem 10 *(Na, K, Cl, CO_2, BUN, creatinine, Mg, Phos)* Pro-BNP/BNP *(until pulmonary hypertension resolved)*
EVERY 2 HOURS (minimum)	ABG *Target ranges for blood gases: pH >7.25, $PaCO_2$ 40-65, PaO_2 60-100*
EVERY 6 HOURS	*Add to ABG:* Hgb iCa Lactate

5. ***Ventilator Management***
    a. Clinical goals:
        i. pH 7.25 – 7.40
        ii. $PaCO_2$ 40-60 mmHg
        iii. PaO_2 > 80 mmHg preductal, > 60 mmHg post ductal
        iv. Pre-ductal SpO_2 >92%
    b. See Appendix for mechanical ventilation titration guidelines
    c. Consider escalation to HFOV if requiring PIP ≥ 28 mmHg, persistent pre-ductal SpO_2 < 92%, $PaCO_2$ > 60 mmHg, 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 ≥ 10
iv. pH 7.28 – 7.4, pCO_2 < 60 mmHg, FiO_2 < 0.6

6. ***Inhaled Nitric Oxide (iNO) (link to iNO protocol)***
    a. ****Patient must have an echocardiogram prior to initiating iNO***
    b. Consider iNO if echocardiographic evidence of PPHN, Oxygenation Index > 20 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. Obtain ECHO within 12 hours of initiating iNO. Then repeat ECHO as needed based upon clinical status.
    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. ***Imaging***
    a. If on CMV, CXR daily to assess for inflation, pneumothorax, ETT placement.
    b. If on HFOV, CXR at least daily but consider twice daily to assess for inflation, pneumothorax.
    c. Obtain baseline HUS if escalation beyond conventional ventilation (in preparation for ECMO if necessary).
    d. Consider obtaining RUS if ECMO anticipated.
9. ***Nutrition***
    a. Initial nutrition should be delivered by TPN. The TPN should not deliver more than 80 cc/kg/day until respiratory/ventilatory parameters and pulmonary vascular pressures are stable. Start on DOL 2. On ECMO, may reduce to 60 cc/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 have been 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. If HFOV or iNO are initiated, order a STAT 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 ≥ 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. $PaO_2 \geq 60$ mmHg
    c. PCO_2 40-45 mmHg
    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 0.4
    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 mmHg, RR 25, or per provider order.
    e. End tidal CO_2 monitoring not indicated until trial off ECMO

APPENDIX

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

OXYGENATION INDEX (OI)

$$\frac{\text{MAP x } FiO_2 \text{ x 100)}}{PaO_2}$$

CONVENTIONAL VENTILATION

Settings/Ranges for CDH Conventional Ventilation Pathway

Mode	Tidal Volume	PEEP	IMV Rate	I-time	FiO2	Pmax
Drager: AC-VG Servo: PRVC	4-5 mL/kg	5 cm H_2O	35-45	0.3-0.45 sec	1.0	28 mmHg

Conventional Ventilator Management Pathway

All ventilator changes should be made only after discussion with the provider

Oxygenation	PEEP	FiO2
SpO_2 < 92%	If FiO_2 1.0, increase PEEP by 1 cm H_2O if CXR reveals under inflation	Increase FiO2 in increments of 5% to max 1.0
$SpO_2 \geq$ 92%	FiO_2 < 0.6, decrease PEEP by 1 cm H_2O if clinically indicated on CXR **Minimum PEEP 5 cm H_2O*	Decrease FiO2 by 5% every hour to maintain goal pre-ductal saturations

HIGH FREQEUNCY OSCILLATOR (HFOV)

Settings/Ranges for CDH HFOV Pathway

Gestational Age	Hertz	Map	Amplitude	Bias Flow
≥ 35 weeks	10	16 (min) – 22 (max) *Use 1-2 above MAP on CMV*	30 (min) – 60 (max)	20

HFOV Management Pathway

All ventilator changes should be made only after discussion with the provider

Oxygenation	Mean Airway Pressure (MAP)	FiO2
SpO_2 < 92%	• If FiO_2 is 1.0, increase MAP by 1-2 every 30 minutes to max of goal range • May need to increase based on discretion of provider and to maintain SpO_2 > 92% • Evaluation of expansion on CXR*	Increase FiO_2 in increments of 5% to max 1.0
SpO_2 ≥ 92%	• FiO_2 < 60%, decrease MAP by 1-2 with scheduled ABGs until minimum range is achieved • Decrease based on ABG results or provider order • Evaluation of expansion on CXR*	Decrease FiO_2 by 5% to goal 0.6 Decrease based on ABG results or provider order

**** Increase / decrease MAP by 1-2 cm H_2O if chest x-ray reveals under expansion (less than 8 ribs visible) or over expansion (greater than 9 ribs visible)***

Patient Assessment/Monitoring

- Chest wiggle: visible bilateral vibration noted from the nipple line to the umbilicus
- Auscultation: changes in the intensity of the piston sounds. (Breath sounds cannot be heard.)
- ABGs: minimum every 2 hours until clinical goal ranges achieved
- Heart rate and infants respiratory rate.
- Transcutaneous CO_2 monitoring
- Oxygen saturation monitoring

Arterial Blood Gas (ABGs)

- Obtain an initial ABG prior to placing patient on HFOV and 1 hour after initiation
- Obtain an ABG 1 hour after any setting adjustments are made
- Obtain ABGs every 2 hours or per provider order

Chest Radiograph (Chest X-ray)

- Baseline chest x-ray if any escalation beyond CMV – goal of 8 rib expansion on HFOV
- 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

- 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 pCO_2, worsening oxygenation (may indicate ETT obstruction, or visible secretions in the airway)

Possible Adverse Effects

- Hyperinflation
- Decreased cardiac output – as evidenced by:
    a. Tachycardia
    b. Decreased peripheral pulse
    c. Decrease in blood pressure
- Pneumothorax – as evidenced by:
    a. Change in ABG
    b. Decrease in oxygen saturations
    c. Decreased chest wiggle on affected side