# The Children's Hospital of Philadelphia
# Newborn/Infant Intensive Care Unit
## *Optimal Care Guidelines*

# *CDH*
## *[Congenital Diaphragmatic Hernia]*

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**I. Early Management**

1. Gentle Ventilation – Permissive Hypercapnia: goal of ventilation is to provide lowest pressures while maintaining adequate lung aeration using a permissive hypercapnia strategy
    a. $PaCO_2$ > 65 acceptable as long as pH > 7.25
    b. Conventional Vent Settings: PIP 20-25 (Goal TV 5 ml/kg), PEEP 5, $t_i$ = 0.3-0.35 depending on rate to maintain minimum I:E 1:2, rate 40 – 80
    c. If $pCO_2$ > 65 at rapid ventilator rate, consider early use of gentle HFOV: Sample settings – MAP 12-14, Hz 6-8. HFOV can be considered as initial strategy in NICU
    d. PIP ≥ 28 cm $H_2O$ or HFOV MAP > 15 used only as a bridge to ECMO
2. Oxygenation
    a. **Primary goal pre-ductal SpO2 > 85%**
    b. **Oxygen titration should be based on pre-ductal saturations. FiO2 should NOT be adjusted based on post-ductal saturations alone.**
        i. Right radial arterial line (pre-ductal), unless echo determination of aberrant right subclavian artery
        ii. UAC and left upper extremity (post-ductal)
    c. Inspired oxygen is started at 50%; weaning begins @ 6 hours of life if stable. Hold weaning of FiO2 at 0.30
    d. Obtain ABG q 1-2h
    e. If shunting > 20% with pre-ductal SpO2 > 85% and PaO2 > 30 with no associated acidosis, team decision must be made about further weaning of FiO2
        i. Shunting:
            1. Ductal shunting: difference between pre and post ductal saturations by 10%
            2. Atrial shunting and/or intrapulmonary shunting: pre and post ductal desaturation with no obvious gradient
            3. Notify FLC/MD if shunting > 20%. Consider trial of increased FiO2 to assess effect
    f. Inhaled Nitric Oxide (iNO)
        i. iNO should be started at 20 PPM
        ii. iNO should be considered independent of the current FiO2 settings in the setting of pre/post ductal pulse saturation difference of > 10 (i.e. pt with a pre-ductal pulse ox of 89% and post-ductal of 75% with FiO2 still at 0.5, iNO could be added at 20 PPM if no other intervention such as BP support or vent support indicated or effective. **FiO2 does not need to be increased when starting iNO.**
        iii. iNO should be considered if pre-ductal pulse ox < 85% and clinical concern of intra-atrial shunting or based on echo findings of right to left atrial shunt.
    g. If persistent PaO2 < 30 and metabolic acidosis, consider need for ECMO.
    h. Place EPIC nursing order with instructions on weaning of FiO2
        1. Wean FIO2 by 3% every hour or 1% every 30 minutes for pre-ductal SpO2 > 85%
        2. Hold weaning of FIO2 at 0.30

3. Do not titrate FiO2 based on PaO2
4. If pre-ductal SpO2 < 85%, increase FiO2 by 5% and notify FLC
5. Notify FLC/MD if shunting > 20%

3. Sedation
   a. Morphine infusion @ 0.01 mg/kg/hr
      i. If hypotensive, use Fentanyl infusion @ 1 mcg/kg/hr
   b. Fentanyl and Versed rescues
      i. Minimize rescues. Increase infusion if necessary
      ii. Avoid Morphine boluses secondary to potential blood pressure effects except as needed for surgical procedures
   c. In general, avoid muscle relaxants
4. Access/Fluids
   a. UAC +/- UVC from initial resuscitation
   b. Peripheral venous lines
   c. PICC line when stable (Lower extremity is preferred) or if UVC unsuccessful
   d. Maintenance fluid of stock D10/D10 + Ca 2 gm/L (in premature infants) + Heparin 1000U/L @ 80 mLs/kg/day
   e. Day 1 start HAL @ 80 mLs/kg/day and continue until start enteral feeds (post repair).
   f. Optimal to remove umbilical lines prior to repair.
5. Blood Pressure Management
   a. Avoid acidosis – goal pH > 7.25
   b. Limit volume – NS boluses in 10 mL/kg increments
   c. Dopamine infusion titrated to support mean arterial blood pressure at or above the patient's gestational age in weeks – max 20 mcg/kg/min
   d. If Dopamine is not available, use Epinephrine infusion @ 0.01 mcg/kg/min
   e. PRBC's (Goal hgb > 12)
      i. Albumin not recommended
   f. Pressor resistant hypotension, strongly consider ECMO consult
   g. Low dose Epinephrine infusion (in rare instances)
6. Medications
   a. Hydrocortisone Sodium Succinate (1mg/kg/dose every 6 hours): 1. order to have at bedside when Dopamine @ 10 mcg/kg/min, start when Dopamine @ 15 mcg/kg/min. 2. Order to have at bedside when Epinephrine @ 0.05 mcg/kg/min, start when Epinephrine @ 0.075 mcg/kg/min.
   b. Dexamethasone (See **Lung Lesion Dexamethasone Administration Protocol)**
      i. Consider in term or near-term infants (> 34 weeks gestation)
         1. Pre-ECMO patients: FiO2 > 80% for > than 48 hrs
         2. ECMO patients: Goal of repair or decannulation in patients that have failed to wean flow, failed trial off, or have CXR with persistent opacification.
         3. NOT intended for use with circuit change
   c. Consider Prostaglandin E1 infusion (0.01 micrograms/kg/min)
      i. Indications for use:
         1. Echo findings (pulmonary hypertension, RV failure)
            a. Consider if there is PH (as a dilator) and/or if PH with RV failure and restrictive/limited PDA. (PH as defined by either systolic and diastolic right to left PDA shunt; TR or IVSP estimate of RVSP => systemic; 2) RV failure is important: BNP> 300; and/or moderate or worse TR; significant R->L atrial shunt)
         2. Persistent metabolic acidosis (pH < 7.25) that is not associated with LV systolic dysfunction by echo
         3. Persistent post ductal PaO2 < 30
         4. May run either peripherally or through CVL, but must run alone
   d. Antibiotics as clinically indicated, not used empirically
7. Studies/Monitoring
   a. Continuous pre and post ductal oxygen saturation monitors
   b. Labs – Type and screen, CBC with diff on admission; BMP/Mg/PO4, bilirubin @ 12 hrs; Lactic acid on admission and q 12 hours x 48 hours.

i. GWA (Genetics lab). Send 1 ml in a sodium heparin/green top tube and 1 ml in an EDTA/lavender top tube. If you can't obtain both just send the EDTA tube. BNP at 24 hours of life, weekly (q Mon), pre PgE1 and 24 hours after starting PgE1, and immediately prior to discharge.

**ii. GWA and Newborn screen must be sent prior to cannulation.**

c. CXR - on arrival, daily and prn
d. Baseline Head Ultrasound on arrival in all cases and repeated as clinically indicated
e. ECHO: 1st as soon as clinically possible on admission. Pertinent findings; ASD, VSD, PDA, LV hypoplasia and Pulmonary Vein Stenosis. 2nd: pre starting PgE1 and 24 hours later; for a clinical question; within 2 weeks of discharge. To order during wqeek day: PH inpatient NP via extend: otherwise the 10154 pager.
    i. Consider peri-op, prior to ECMO decannulation, acute change in BNP, prior to and during weaning of NO, post NO, and prn per PH team.

8. EKG technician to obtain EKG, order through EPIC so that it can be read by Cardiology and available in MUSE
    a. Optimal timing of EKG: pre-op

**II. Pulmonary Hypertension**

1. Best therapy is prevention
    a. Common triggers for PH crises – suctioning ETT, stimulation of patient, decreased pH, increased $pCO_{2\ (>65)}$, and hypoxemia
2. Consult Pulmonary Hypertension Team. Ensure that call was made to PH team if echo was performed by cardiology consult team.
3. ECHO:
    a. Consider pertinent findings: TR, intraventricular septal position, presence or absence of PFO, presence or absence of PDA (PDA size?), direction of shunt (both at PFO and PDA: systolic and diastolic), RV/LV dysfunction
4. Inhaled Nitric Oxide (iNO) and FiO2
    a. Continue iNO until BNP < 100, right ventricular pressure < systemic blood pressure after CDH repair or FiO2 consistently < 0.30
    b. Consider weaning FIO2 below 0.30 once off iNO and BNP < 100
    c. Discontinue iNO and keep FiO2 < 0.30 while on ECMO and reintroduce pre-decannulation and lungs aerated
5. Sildenafil: as per consultation with PH Cardiology
6. Consider post pyloric feedings if there is any unexplained inability to wean the FiO2, iNO, or ventilator

**III. ECMO**

1. Indications
    a. Inability to maintain pre ductal saturations > 85% or post ductal $PaO_2$ > 30
    b. PIP > 28 mm Hg, MAP > 15
    c. Pressor resistant hypotension
    d. Inadequate oxygen delivery based on persistent metabolic acidosis or rising serum lactate level
    e. Inability to wean from $FiO_2$ 100% in first 48 hours of life
2. Inclusion Criteria
    a. BW > 2 kg
    b. GA > 34 wks
    c. Absence of Intracranial hemorrhage > Grade I
    d. Absence of other significant congenital or chromosomal anomalies
    e. Absence of significant cardiac defects (HLHS, coarctation,etc)
3. ECMO Considerations
    a. GWA and Newborn screen must be sent prior to cannulation
    b. Send ECMO coagulation panel (PT, PTT, INR, Fibrinogen, AT3, D-Dimer, ACT), BNP
    c. Review current ECMO Anticoagulation Guidelines
    d. Recommend ordering blood products for ECMO prime first followed by call to blood bank
4. ECMO Parameters

a. Rest settings: FiO2 0.30, PIP 20, PEEP 10, $t_i$ 1.0, rate 10, PS 10 x 2-3 days and then consider adjustment of settings (i.e. decrease peep and $t_i$)
b. Goal $PaCO_2$ 45-55
c. Diuresis with Lasix infusion starting @ 0.04mg/kg/hour and titrating to effect beginning ECMO day 2-3 once flow issues stabilized. Follow BUN, Cr, base excess. Consider scuff of blood products after day 3.
d. Head Ultrasound (daily x 5, then every other day)

5. Discontinuation of ECMO
   a. Consider ETT change in 24 hours prior to planned decannulation
   b. Coordinate time for trial off with ECMO team, nursing, surgery, and neonatology
   c. Determine appropriate vent settings and plan for trial off with clinical team
   d. Arrange for ECHO to assess pulmonary pressures during trial off . (This requires an attending ECHO cardiologist, so appropriate notice is required)
   e. Monitor tCOM and saturations continuously during ECMO flow wean
   f. Monitor blood gases q hour during wean and adjust vent accordingly
   g. Equipment for decannulation should be at bedside (Happy Pack, broviac and ECMO decannulation tray)
   h. Hold HAL and replace with stock (as per current peri-op guidelines) for decannulation to avoid problems with potassium
   i. OR team scheduled for selected difficult cases
   j. Trial off successful when adequate oxygenation and ventilation (ideally $FIO_2 < 50\%$, PIP < 28, MAP < 12)

**IV. Surgical Repair CDH**

1. Timing - Delayed until on $FIO_2 < 50\%$, PIP < 25 cm $H_2O$, MAP < 12, resolution of pulmonary hypertension (Clinical criteria, consider echo), normal acid base balance, stable blood pressure, resolution of anasarca, nearly ready to come off ECMO or already off ECMO
2. Location – nearly all cases will be done in the NICU, on the baby's warmer bed
3. Scheduling – These cases must be scheduled early in the day to ensure adequate support for patient care both intra-operatively and post-operatively

4. Preparation
   a. If patient is on ECMO, consider use of AMICAR infusion @ 20 mg/kg/hr following load of 100 mg/kg. Review massive hemorrhage protocol found on share drive:share.chop.edu under Nicudocs/Division of Neonatology/Guidelines, Policies, & Protocols
   b. Order blood products (to be in OR or NICU at bedside during procedure). Blood products will be drawn up in syringes to allow rapid infusion
   c. Clear space for access by anesthesia and surgery – all nonessential equipment and people removed
   d. Ensure adequate venous access for administration of blood products, medications. Umbilical venous line will be removed
   e. Ensure adequate arterial access for monitoring of blood pressure and blood gases. Peripheral arterial access is optimal
   f. Hold HAL and replace with stock (as per current peri-op guidelines) for surgery to avoid problems with potassium
   g. Administer preoperative antibiotics
   h. Bear Hugger, Bovie, and Surgeon's Headlight at bedside, trans-warmer
5. Monitoring
   a. Pulse oximetry monitors, both pre and post ductal must be in full view
   b. Blood gases will be monitored at baseline and then approximately every 30 minutes
   c. In addition to surgical team, attending or fellow neonatologist, bedside nurse, respiratory therapist should be present. Communication between anesthesia, surgery, and neonatology is imperative.

**V. Discharge and Follow Up**

1. Mid-Hospitalization:
    a. PFT's
        i. Consider inpatient consult with Dr. Panitch for PHP patients > 30 days old who have oxygen requirement or CLD. (May require PFT's prior to discharge)
2. Discharge
    a. MRI
        i. Brain MRI/MRA "without contrast" with susceptibility for ALL patient's post ECMO.
        ii. Brain MRI "without contrast" with susceptibility for ALL non ECMO
    b. Audiology
        i. ABR/OAE screening
        ii. Audiology follow-up should be recommended for 6 months of age.
    c. Immunizations
        i. Will need Synagis monthly during RSV seasons
        ii. Flu vaccine if greater than 6 months
        iii. Generally, no contraindications for administration of Rototeq
3. PHP Follow-Up
    a. Consider baseline CXR prior to discharge. Consider copy of CXR for parents.
    b. Schedule standard 2 week follow-up appt with primary surgeon.
    c. Schedule standard 1 month follow up with Cardiology
    d. PHP appointment intervals as follows: 6 months, 1 year, 2 year, 4½ -5 years (Pre K), 6 year, then every other year.