Congenital Diaphragmatic Hernia Consensus Guidelines

August 2017

I. **Delivery Plan (Referring L&D Hospital)**

Planning Before the Delivery Occurs:

- Antenatal steroid given as indicated for preterm labor/OB indication.
- Delivery route and timing determined by OB/Perinatology/Parents.
- Request placenta and cord gasses
- Delivery to occur in largest Delivery Room (D)
- Team Captain for Neonatal Resuscitation is the neonatal attending on-service/on-call at referring L&D hospital.
- Attending should be notified to be present for delivery.
- Team Captain reviews each team member's role with team*
    - **Resuscitation Team:**
        - Team Captain (referring L&D hospital NICU Attending who is on-service/on-call)
            - Runs resuscitation, available for intubation. Intubation will be by most experienced provider, at discretion of Team Captain
        - Assisting Clinician
            - Ready to insert line under sterile conditions.
            - Places low UVC
        - Two Respiratory Therapists, including one experienced Respiratory Therapist who is comfortable and efficient with intubations.
            - More experienced RT to intubate or assist with intubation (at discretion of team captain).
            - Less experienced RT places preductal pulse oximeter, runs blood gases and assists with securing ETT
            - Two NICU Nurses
                - One nurse documents
                - Other nurse applies restraints, administers medications and places replogle and connects to suction.

Checklist for Physicians:

NICU Patient Bedside Prep

- € Designate bed space for patient in consultation with Charge RN
- € Request bedside set up with:
    - Double suction canister set up [1 for respiratory suction, 1 for Replogle]
    - Pre-post ductal saturation monitoring.
    - IV pumps for UAC/UVC maintenance fluids and two more pumps for sedation, minimum.
    - Conventional ventilator at bedside
    - HFOV calibrated and on stand-by
- € Prepare umbilical line set up (non-DR lines), primed with heparinized saline and cover with sterile towels.

DR Bedside Prep (Use NICU emergency cart located in OR area)

- € Prepare umbilical line set up for DR, single lumen UVC primed with heparinized saline and cover with sterile towels.
- € Place covered prep in OR area and bring cart into DR when team is called to delivery

Pre-admission Orders (prepare on handwritten MD order form)

- € IVF
  - UVC or PIV maintenance: D10W with Calcium gluconate 200mg/kg/d and heparin 0.5 Unit/mL IVF to run at 80mL/kg/d
  - UAC: 0.9%NaAcetate + Heparin 0.5 unit/mL to run at 1ml/hr
  - Fentanyl drip 10mcg/mL, starting dose 1mcg/kg/hour
  - Dopamine drip 1600mcg/mL, starting dose 5mcg/kg/min
  - Epinephrine drip 100mcg/mL, starting dose 0.05mcg/kg/min

- € Delivery Room Medications to Have Available
  - Fentanyl 1 mcg/kg IV, Two doses drawn up
  - Versed 0.1mg/kg IV, Two doses drawn up
  - Norcuron 0.1mg/kg IV, Two doses drawn up
  - Normal Saline 10ml/kg, Two doses drawn up
  - THAM (if available) 5ml/kg, Two doses drawn up
  - NS flush 3ml, 2-5 Doses in single dose syringes

Checklist for Respiratory Therapists: Preparation should be completed prior to induction or at least 2 hours before scheduled C-section.

- € Laryngoscope/Blade
- € ETT/stylet
- € PediCap
- € ETT Tape/Neobar
- € Pulse oximeter
- € Mask
- € Suction catheter (8 and 6 Fr)
- € Prepare Conventional Mechanical Ventilator and High Frequency Oscillator should be calibrated and available on stand-by.

Checklist for Nurses: Preparation should be completed prior to induction or at least 2 hours before scheduled C-section.

- € Soft Restraints (Four)
- € Replogle, 10Fr and 8Fr (for Birthweight <1.5 kg)
- € UVC Tray (includes Scalpel, Umbilical Stump Tie, Betadine)
- € Op-Site
- € Alcohol swabs
- € Fentanyl 1 mcg/kg for slow push (Two Doses drawn up)
- € Versed 0.1 mg/kg bolus (Two Doses drawn up)
- € Norcuron 0.1mg/kg IV, (Two Doses drawn up in single dose syringes
- € NS bolus 10 ml/kg (Two Doses drawn up)
- € THAM (if available)5 ml/kg bolus (Two Doses drawn up)
- € Flush for meds and clearing lines
- € 20G Angiocath (Two) and 23 G Butterfly needle (Two) with T-connector and stopcock and syringes for pneumothorax
- € 10ml syringe (Three)
- € 1ml syringe with heparin (Four)
- € DR Warmer turned on and warming

II. Delivery Room Management/ Initial Stabilization

- Infant immediately handed to neonatology (no delayed cord clamping)
- Cord blood (artery and vein) drawn by OB team for blood gas analysis.
    - Occurs simultaneously within first two minutes:
        - Minimal handling until baby is intubated
        - Designated MD/RT intubates and verifies ETT position with PediCap
            - Positive Pressure Instructions: Initial maximum PIP 20 for three breaths with long I-time (if need more pressure, then may increase up to PIP 25 for three breaths, then adjust (PIP<25) based on compliance
            - Start with FiO2 1.0
            - Initial pre-ductal oxygen saturations >65% acceptable
            - After first three breaths, respiratory rate 40-60 with IT 0.4 or shorter if using rate closer to 60.
        - Other RT places pre-ductal pulse oximeter
        - RTs secure ETT
        - Nurse places replogle and connects to low continuous suction
        - Nurse applies soft restraints
        - Assisting clinician places low UVC and secures with Op-Site
        - Nurse gives Fentanyl over 10 minutes on a pump (Versed/Norcuron only for complicated resuscitation, severe agitation, per Attending MD request)
    - Goals:
        - HR>100, pre-ductal saturations>65% initially
        - By 5 minutes, pre-ductal saturations >70%
    - Transport to NICU when stabilized, ideally by 10 minutes, on DR warmer

- <u>In NICU at referring L&D hospital</u>:
    - Conventional Mechanical Ventilation is the preferred mode. Place patient on: Rate 40–60, PIP up to 25, PEEP 5-6, IT 0.4 seconds (may need shorter if using higher rate), FiO2 100%.

    - Goal pre-ductal saturation 80-95%
    - Target PCO2 between 50-70 with pH>7.25
    - Consider HFOV if unable to oxygenate or ventilate effectively despite maximizing conventional support
    - If pre-ductal saturations >95% may slowly titrate FiO2 downward; hold if evidence of lability.
    - Obtain blood gas by 30 minutes of life
- Place UAC/UVC (confirmation of placement of UVC by ECHO after infant transferred to CHLA)
    - Goal is adequate perfusion with blood pressure within normal levels for gestational age (MAP 40-50 for term infant)
    - For signs of poor perfusion/hypovolemia, volume bolus of 10mL/kg may be attempted, up to 3 times.
    - Dopamine is first line agent for pressor support.
    - Consider addition of Hydrocortisone for pressor resistant hypotension.
    - Epinephrine may be considered as second line agent.
    - Ensure normal ionized calcium level.
- Initiate D10W with Calcium gluconate 200mg/kg/d and heparin 0.5 Unit/mL IVF to keep total fluids at 80mL/kg/d (total fluids) via PIV/UVC
- Initiate 0.9%NaAcetate + Heparin 0.5 unit/mL to run at 1ml/hr via UAC
- Fentanyl infusion should be initiated at 1 mcg/kg/hour

III. Medical Management in the NICCU

<u>Ventilation Management</u>:

- Conventional Mechanical Ventilation is the preferred mode of ventilation. Consider HFOV if unable to ventilate with acceptable setting on CMV
- Place patient on: Rate 40–60, PIP up to 25, PEEP 5-6, IT 0.4 (or less if using higher rate). If HFOV utilized, limit MAP to <18, 8-10Hz, amplitude 30-50. Consider HFOV if unable to maintain pCO2 in target range with PIP <25, Tidal Volumes 3-5 mL/kg and/or unable to reach preductal saturations >85%.
- Adapt treatment to reach a preductal saturation >85% and post-ductal saturation >70%
    - FiO2 should be slowly weaned (3-5% every 4-6 hours) when preductal saturation remains >95% and post-ductal saturation >92% without significant lability. Faster wean (3-5% every 1-2 hours) should be utilized if saturations persistently >98% and paO2 persistently >80.
- Target PaCO2 range should be between 50-70 mmHg, with weaning when pCO2 <50
- Target pH 7.30-7.40, >7.20 may be acceptable if PCO2 in range and adequate tissue perfusion, Lactate <20
    - *Lability of patient oxygenation and hemodynamics must be considered and adjustments to target pH/CO2/pace of weaning may need to be altered based on this lability.

- Presence of air-leak: pneumothorax should be evacuated by placement of thoracostomy tube. If pneumothorax is on side of the hernia, surgical fellow should be consulted for placement of

tube by surgical fellow or attending. HFOV/HFJV should be ventilator of choice in presence of air-leak.

- Monitoring:
    - X-rays, upon admission, after line/tube placement, after change in ventilator mode.
    - Otherwise daily on CMV, twice a day on HFOV, and with significant clinical deterioration.
    - Transcutaneous CO2 monitoring, pre and post-ductal saturation monitoring continuously.
    - Blood gasses q4-6 hours initially, spacing dependent on stability.

Hemodynamic Management:

- Goal is adequate perfusion with blood pressure within normal levels for gestational age (MAP 40-50 for term infant).
- Effects of and choice of treatment may best be guided by repeat echocardiographic evaluation.
- For signs of poor perfusion/hypovolemia, volume bolus of 10cc/kg may be attempted, but should typically be limited to < 40mL/kg total boluses (including at birth hospital) prior to additional therapies.
- Dopamine is generally the first line agent for inotropic support
- Consider addition of Hydrocortisone for pressor resistant hypotension
    - Send cortisol level at 24 hours to screen for risk of pressor resistant hypotension and assist with weaning once off pressor support.
    - If considering hydrocortisone prior to 24 hours of life, send cortisol level prior to administration for assistance with weaning once off pressor support
- Epinephrine may be considered as second line inotrope
- Additional agents that may be considered based on physiology/echocardiographic evaluation: Milrinone, Vasopressin, Norepinepherine.

Pulmonary Hypertension Management:

- Perform echocardiography upon admission to the NICCU
- Blood pressure support should be provided to maintain arterial blood pressure levels at normal levels for gestational age
- iNO should be considered once lungs are recruited if evidence of elevated TR jet or right to left shunting with saturation difference >10%
- Sildenafil may be considered as a second line agent for significant pulmonary hypertension by echo.
- Milrinone may be considered for pulmonary hypertension, but may worsen hypotension related to afterload reduction and should be utilized with caution.
- Flolan may be considered in patients who are not ECMO candidates or would be considered high risk ECMO candidates.
- Pulmonary vasodilators with more potential systemic effects should be weaned prior to weaning iNO (i.e wean Flolan, then milrinone, then sildenafil).

Vascular Access:

- Appropriate position of UAC/UVC must be confirmed upon admission to CHLA
- UVC position must be confirmed by echo given difficulty confirming non-hepatic positioning in presence of liver herniation as well as mediastinal shift

- Alternate central line should be placed within 3-6 hours if unable to place or confirm appropriate placement of UVC.
- Placement of a right radial PAL to monitor pre-ductal gasses should be considered if UAC unable to be placed, is malposition, or arterial monitoring is indicated beyond 7 days.

ECMO Criteria: ECMO should be considered in the setting of any or all of the following physiological criteria after attending level discussion between neonatology and surgery.

- Inability to maintain preductal saturations>85% despite maximal medical management
- Increased PaCO2 and persistent respiratory acidosis <7.20 with need for PIP >25, MAP>18
- Inadequate oxygen delivery with post-ductal paO2<40, increasing lactate>30, or metabolicacidosis with pH<7.15.
- Oxygenation index>40 repeated x 3, A-a Gradient >600-624.
- Systemic hypotension resistant to fluid and inotropic therapy.
- Absence of contraindications (presence of relative contraindications should be discussed by multidisciplinary team).

ECMO Management:

- Discussion between neonatologist and surgeon, re: VA vs VV. VA is most commonly utilized, but VV may be considered based on primary indication for ECMO and cardiac function.
- Goal to maintain lung recruitment while minimizing barotrauma, typically with conventional ventilation, high PEEP strategy (i.e. 20-22/10-12 with rate of 10-20, IT 0.8-1)
    - Settings should be decreased judiciously post-cannulation, following transcutaneous CO2, blood gasses, saturations, and serial CXR's
    - VA: FiO2 should be weaned to 30%; VV: FiO2 should be weaned slowly as tolerated to maintain goals
    - In presence of airleak, may consider HFOV for rest settings
- VA: Flow should be targeted to maintain SVO2 75-85, preductal saturations >95%, paO2 >60
- VV: Flow should be targeted to maintain preductal saturations>85%
- During weaning phase and trial off ECMO, ventilator settings should be adjusted to be consistent with ventilation management guidelines above
- iNO may be weaned off once on ECMO if lungs are collapsed, with consideration of resuming later in course if lungs recruited and evidence of significant pulmonary hypertension.
- If lung recruitment maintained, may consider continuing iNO in setting of persistent moderate to severe pulmonary hypertension
- Management of additional pulmonary vasodilators should be directed by echocardiographic reassessment as well as assessment of lung recruitment/fluid balance.
    - Decisions related to pulmonary hypertension management/hemodynamics should ideally be made with echocardiographic assessment on minimal flow/during trial off, although trends may be monitored by echocardiograms on full support
- See fluid/neuro sections regarding details around fluid/electrolyte management (including use of CVVH) and neurology consultation
- See surgical management for details regarding timing of repair
- Readiness for decannulation should be discussed based on ability to wean flows, ventilator and oxygen requirements during weaning/trial off, improvement in or resolution of pulmonary hypertension and cardiac dysfunction, return to dry-weight, and de-escalation of medical therapy for pulmonary hypertension.

- Progress and readiness for trial off/decannulation should be discussed daily between the neonatology and surgery teams and will also be dependent upon timing of repair, complications, and prognosis.

Fluid Management and Parenteral Feeding:

- Fluids should be provided judiciously with goal to maintain intravascular volume status while avoiding significant edema/lung water. Initial fluid management should be restrictive (60-80 mL/kg/d) with emphasis on maximizing nutrient and glucose delivery while minimizing fluid volume.
- Once central access is secured, fluid should contain 400mg/kg/d Ca-Gluconate. Specific attention to electrolyte balance should be paid, particularly Calcium and Magnesium
- Nutrition should be optimized to provide adequate calories to meet metabolic demands
- Diuretics may be considered after first 24-48 hours, in case of persistent positive fluid balance (if not on ECMO).
- CVVH should be initiated for all patients requiring ECMO, with goal of achieving neutral balance within the first 24 hours, followed by initiation of fluid removal as tolerated to achieve negative balance. Goal is to achieve negative fluid balance with return to "dry weight" to facilitate weaning and minimize lung water.

Gastrointestinal Management

- Replogle to low intermittent suction pre-repair
- Careful attention to management of gaseous distension on serial x-rays
- H2 blocker should be added to TPN and continued when enteral feeds initiated as part of reflux management
- NPO until return to bowel function post-operatively
- Balanced nutrition with adequate kcals/kg should be provided via TPN prior to initiation of feeds
- Advancement of feedings after tolerance of trophic feeds for 3-5 days, with typical advancement by no more than 20/kg/d.

Neurologic Management

- Maintenance of a low stimulation environment: ear plugs, low light, minimal stimulation, limiting "touch times" to q6 hours, no baths.
- Sedation for comfort and to minimize gaseous distension, while minimizing hemodynamic effects of narcotics/sedatives
    - Fentanyl continuous drip, titrated to effect
    - Benzodiazepines may be considered with significant agitation unresponsive to comfort measures and Fentanyl gtt, but hemodynamic effects must be considered as well as potential neurologic sequelae
    - Consideration of low dose Dexametomodine given potentially less hemodynamic effects, in consultation with Pain Team.
- Use of neuromuscular blockade should be minimized
    - Use should be approved by Attending Neonatologist
    - If utilized, necessity should be reassessed every 6 hours

- Cranial Ultrasound upon admission and within 12 hours of ECMO (if necessary) and repeated per ECMO guidelines if requiring ECMO support
- Consideration of NIRS monitoring for identifying trends
- CFM and Neurology consultation for all ECMO patients (will typically recommend full video EEG for first 24 hours)
- CFM should be utilized if patient requiring neuromuscular blockade

Genetics

- Prenatal ultrasound results and any genetic testing performed should be reviewed by the clinical team upon admission.
- Postnatal genetics consultation should be requested for any infant with CDH plus any dysmorphic features, second major congenital malformation, IUGR/SGA, abnormal prenatal genetic testing.
- Postnatal genetic consultation should be considered in apparent isolated CDH given subtlety of many dysmorphic features.
- Strongly consider obtaining chromosomal microarray prior to ECMO cannulation/blood transfusion as well as obtaining DNA prior to demise for possible future testing, in consultation with Genetic service.

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