

NICU Congenital Diaphragmatic Hernia (CDH) Care Guideline



Delivery Room Management

- Consider 30 seconds delayed cord clamping.
- Place infant on Giraffe warmer.
- Immediate intubation and place Pre- and post-ductal saturation monitors.
- Place 10Fr salem sump OG or NG Tube to low continuous suction (LCS).
- Obtain Microarray from cord blood.
- If unable to attain 80% saturations in the delivery room using CDH ventilation strategies, notify NICU to have HFOV at bedside.

NICU Admission

- Admit to 4S NICU, notify Red Team attending/fellow.
- Verify 10Fr salem sump is in position.
- Insert UVC and UAC - placement verified with CXR/KUB xray.
- Admission Labs: Transport Work-up, Blood Culture, CBC, q4h Blood Gases with Lactate
- Notify Pediatric Surgery, ECMO On-Call Physician.
- Pre- and post-ductal saturation monitoring.
- Administer Erythromycin and Vitamin K per orders.
- Initiate sedation early (Dexmedetomidine gtt or intermittent narcotic doses) {see CDH Sedation Protocol Box}.
- Cranial Ultrasound.
- Immediate echocardiogram with severe defects or within 12 - 24 hours of admission, for mild defects.
- Consider the use of antibiotics based on risk factors for perinatal infection. Does not require empiric antibiotic coverage.

NICU Management

- Gentle ventilation (See CDH Ventilation Strategies).
- If unable to achieve targets, notify Red Team on-call, may consider HFOV.
- If non-central UVC, obtain PICC within 24 hours.
- Maintain total fluid goal at 60 - 100 ml/kg/day.
- Initiate Dexmedetomidine gtt and maintain adequate sedation (see inset).
- Goal Mean Arterial Pressure (MAP): 40 - 50 mmHg. If low, identify the ideology of hypotension and treat accordingly, consider Epinephrine IV 0.02 mcg/kg/min.
- 24 Hour of Life Labs: Neo CMP, Newborn Screen, CBC

Pre-Op Work-up and Criteria for Repair

- Schedule repair when physiological stability has been demonstrated.
- Order 2 units PRBCs, Platelets, Fresh Frozen Plasma on-hold for OR.
- Ensure abdominal US performed prior to CDH repair.
- Pre-operative labs: CBC w/diff, CMP, Type and Cross, DIC.
- Obtain CXR and Blood Gas prior to OR.
- cNIRS with goal targets > 50
- Pain/Sedation – [Refer to NICU Pain Guidelines](#)
- IV Cefazolin x 24 hours for perioperative antibiotic prophylaxis:
 - ≤ 7 days and < 2000g – 25 mg/kg q12h
 - > 7days and < 2000g – 25 mg/kg q8h
 - > 7 days and > 2000g – 50 mg/kg q8h

Post-Operative Care

- Post-operative labs: CMP, CBC, DIC x 1 post-op
 - Blood Gas with Lactate q4 hours x 24 hours post-op
- Vital Signs per policy, monitor for signs of abdominal compartment syndrome (e.g. - hypotension, oliguria, abdominal distension, difficulty ventilating/breathing)
- Goal: Pre-ductal saturations > 90%.
- Remain NPO on POD #0 with 10Fr salem sump to LCS.

Hyperlinks

- **Refer to:** [UTHealth – Management of Infants with Congenital Diaphragmatic Hernia from Birth to Surgery](#)
- **Refer to:** [ELSO Guidelines – Management of Congenital Diaphragmatic Hernia Treated with Extracorporeal Life Support: Interim Guidelines Consensus Statement From the Extracorporeal Life Support Organization](#)

Physiologic Targets

Pre-ductal Saturations: > 80 - 85%
Post-ductal Saturations: > 75%
pH: > 7.2
PaCO₂: 50 - 70 mmHg
PaO₂: 40 - 90 mmHg

CDH Ventilation Strategies

- Mode: SIMV/PC or SIMV/VG per physician discretion
- PIP: 20 cmH₂O (Max 26 cmH₂O) *Target Volumes 4 – 5 cc/kg*
- PEEP: 5 cmH₂O (Max 6 cmH₂O)
- Rate: 40 bpm (Max 50 bpm)
- iTime: 0.3 seconds
- FiO₂: Initiate at 50% (Titrate to pre-ductal saturations 80%)

CDH HFOV Settings

- Max MAP: 10 - 15 mmHg
- Amps: 25 - 40 Hz (Target: perceptible “jiggle”)
- Hz: 8 - 15

Additional Therapies

- Inhaled Nitric Oxide (iNO) and/or Prostaglandin (PGE) may be used for pulmonary hypertension (pHTN) only after ECHO and discussion with NICU Red Team.
- iNO is contra-indicated in infants with left ventricular (LV) dysfunction, left atrial (LA) enlargement.
- PGE may benefit infants with pHTN and right ventricular (RV) or biventricular cardiac dysfunction .
- Early use of IV Hydrocortisone for refractory hypotension should be utilized {see dosing on Page 3}

CDH Sedation Protocol

First Line: Dexmedetomidine gtt – start 0.3 mcg/kg/hr (max dose 1.5 mcg/kg/hr) Note: Avoid HR < 100 and limit use to 5 - 7 days.

Second Line: Initiate Morphine 0.05 mg/kg/hr.

- Vecuronium: Consider as needed to minimize oxygen demand and in cases of physiologic instability.

Extracorporeal Life Support (ECMO)

Criteria: *O₂* > 40, *PaO₂*: persistently < 40, *Lactate*: persistently > 3 mmol/L, Unable to obtain physiologic targets on maximum ventilatory support or unreasonable vasoactive support.

Relative Contraindications to ECMO

- GA ≤ 34 weeks and weight ≤ 1.7 – 2 kg
- Concomitant severe Congenital Heart Disease after discussion with multi-disciplinary team.
- Major genetic abnormalities and additional major, severe surgical anomalies.

Additional Resources

Page 2: Prenatal & Delivery Room Recommendations
Pages 3-5: NICU Interventions
Page 6: Postop Care and Discharge Criteria

Inclusion Criteria: All infants born with congenital diaphragmatic hernia

Available Resources:

- CDH Parent/Family Education

Prenatal Recommendations

Antepartum Care:

- Ultrasound suspicious for CDH – refer to Maternal Fetal Medicine (MFM) for detailed ultrasound exam.
 - MFM ultrasound to include evaluation for other abnormalities - Lung area to head circumference ratio (LHR) and observed to expected (O/E) ratios, description of liver involvement, and preliminary counseling/consultation.
 - May also consider fetal MRI.
- Referral to Pediatric Cardiology for fetal echocardiogram at approximately 22 weeks.
- Genetics consultation with discussion of amniocentesis for microarray.
- Referral to Pediatric Surgery.
- Ongoing fetal surveillance to include ultrasound approximately every 2 weeks to evaluate fetal status, fluid and grow as appropriate.
- Initiation of antepartum fetal monitoring with twice weekly Nonstress test (NST) / weekly Amniotic fluid index (AFI) at 33 - 34 weeks or sooner if other co-morbidities (for example, intrauterine growth restriction (IUGR) are noted.
- Multidisciplinary care meeting to involve Obstetrics (OB), MFM, Neonatology, Genetics and Pediatric Surgery.

Delivery:

- Recommended delivery at a tertiary medical center with ECMO capabilities.
- Routine caesarean section is *not* recommended.
- Discussion of possibility of induction of labor at fetal consultation.
- Encouragement of full term delivery (39 weeks) but delivery may be warranted earlier for fetal and/ or maternal indications.
- Use of antenatal steroids may be indicated in infants less than 37 weeks gestation with concern for preterm delivery.

Delivery Room Anticipation and Resuscitation

Pre-briefing:

- Team huddle with discussion of plan of care and clearly defined team member roles.
- Advanced preparation of supplies including equipment for intubation, 10Fr salem sump, potential normal saline fluid boluses and resuscitative medications. Have CODE cart immediately available.

Delivery / Resuscitation:

- Consider delayed cord clamping \approx 30 seconds.
- No resuscitation with positive pressure by mask, if possible.
- Immediate intubation
 - Start at rate of 40 bpm (Max 50 bpm)
 - Peak Inspiratory Pressure (PIP) 20 cmH₂O with **Target Volumes 4-5 cc/kg**
 - Adjust PIP to max 26 cmH₂O.
 - Positive end-expiratory pressure (PEEP) 5 cmH₂O (Max 6 cmH₂O)
 - Inspiratory Time (IT) 0.3 seconds
- Placement of 10Fr salem sump orogastric or nasogastric tube to low continuous suction.
- Placement of pre-ductal pulse oximeter.

Initial Saturation and Oxygen Use Goals:

- Inspired oxygen is started at 50%.
- Pre-ductal saturations greater than 65% at 5 minutes of life; greater than 75% at 10 minutes of life.
- In the event the infant remains bradycardic, may increase the oxygen concentration to 100%.

Neonatal Intensive Care Interventions

Monitoring:

- NICU monitors with pre- and post-ductal saturations.
- Transcutaneous oximetry measurement (Tcom) and end-tidal monitoring (ETCO₂).
- May place NIRS cerebral monitoring.

Diagnostic Studies/Labs:

- Admission labs: Transport work up, blood culture, CBC, Microarray.
- CXR on admission and as indicated.
- Baseline head ultrasound on arrival.
- Echocardiogram on admission.
- Ultrasound of chest, abdomen, CDH.
- Consults: Pediatric Surgery, Genetics.

Ventilation:

- Goal of gentle ventilation by means of permissive hypercapnia to provide low pressures while maintaining adequate lung aeration.
 - Acceptable PaCO₂ range: 50 – 70 mmHg
 - Acceptable PaO₂ range: > 40 mmHg
 - Acceptable pH range: 7.20 – 7.40
 - Lactate goal: < 3 mmol/L
 - Ventilator settings:
 - SIMV preferred
 - Rate: 30 – 40 bpm
 - PIP not to exceed 26 cmH₂O
 - iTime: 0.3 seconds
 - Goal Tidal Volume: 4 – 5 cc/kg
- Consider the use of high frequency oscillatory ventilation (HFOV) if increased settings are required. MAP not to exceed > 15 mmHg unless used briefly as temporizing measure. Consult ECMO team if transitioning to HFOV.

Oxygenation:

- Pre-ductal saturation goal: > 80 - 85%
- *Oxygen is started at 50% - weaning can begin at 6 hours of life, if stable. Hold weaning at 30% until infant has demonstrated stability and consistent high oxygen saturations.*
 - *Wean O₂ by 3% every hour for pre-ductal saturations greater than 85%.*
 - *If pre-ductal saturations are less than 85%, increase O₂ gradually as needed, consider echocardiography.*
- Surfactant: The routine use of surfactant has not been demonstrated to improve outcomes in CDH. However, in infants born prematurely, particularly those who did not receive antenatal steroids, surfactant can be considered on a case-by-case basis.
- Initiation of Inhaled Nitric Oxide (iNO) and indications of use: (see also considerations for management section for more information).
 - Echocardiogram demonstration of pulmonary hypertension that is unresponsive to other management options and infant is requiring greater than 60% oxygen with *NO* evidence of isolated, left-heart dysfunction.
 - NICU Red Team and ECMO and Surgical teams should be made aware of any patient requiring initiation of iNO.

IV Access / Fluid Management:

- UAC/UVC or peripheral arterial line and PICC line.
 - Placement of umbilical venous and arterial lines: if liver is in thoracic cavity, the UV catheter often does not travel thru the ductus venous and alternative venous access should be obtained when patient is stable.
- Maintenance fluid ≤ 60 ml/kg/day including all current infusion.

Continued Neonatal Intensive Care Interventions

Hemodynamic Management:

- Target goal: Mean Arterial Pressure of 40 – 50 mmHg with adequate urine output and lactate < 3 mmol/L. In some cases, higher MAPs may temporarily improve oxygenation; however, targeting supra-physiologic blood pressures to “reverse” the ductal shunting is not recommended for long periods as this can compromise systemic blood flow.
- Initiation of infusions:
 - Consider echocardiogram for all unstable infants on admission to help guide hemodynamic management. Otherwise, obtain echocardiogram within 24 hours of life, or with onset of clinical instability/refractory hypotension.
 - If unable to obtain echocardiogram or if hypotensive and awaiting imaging, consider Epinephrine as front line (Initiation at 0.02 mcg/kg/min for inotropy. If requiring escalation above 0.10 mcg/kg/min, may need to consider stress dose of hydrocortisone and addition of 2nd line agent).
 - Consider early stress dose hydrocortisone dosing (50 mg/m²) followed by 1 mg/kg q6hours for refractory hypotension.
 - Norepinephrine/Vasopressin can be utilized as second line agents after speaking with Red Team attending and Surgical team.
 - Dopamine should be avoided in cases of moderate to severe pulmonary hypertension or cardiac dysfunction.
 - All infants with CDH should undergo echocardiogram within 24 hours of admission. Consider stat echo if refractory hypotension or escalating towards ECMO.
 - Consult ECMO team in the event of continued hypotension.

Antibiotics:

- Consider the use of antibiotics based on risk factors for perinatal infection. Does not require empiric antibiotic coverage.
 - Can consider stopping at 36 hours if labs and clinical stability are reassuring.

Sedation:

- As clinically indicated
 - Use of intermittent Morphine dosing first
 - Dexmedetomidine – Use as first line in most infants unless significant agitation leading to instability. Start 0.3 mcg/kg/hr (max dose 1.5 mcg/kg/hr) – limit use to 5-7 days.
 - Morphine or Fentanyl infusion – Second line, or first line in infants with significant instability/hemodynamic compromise, IV Midazolam PRN if indicated.
 - While the routine use of Vecuronium for muscle paralysis is not recommended, in infants with severe hypoxemic respiratory failure or evidence of compromised oxygen delivery (lactate elevation, down trending cerebral/renal NIRS, mixed venous saturation < 50), neuromuscular blockade may assist with ability to oxygenate/ventilate.

Pulmonary Hypertension:

- Prevention is best.
- ECHO finding suggestive of systemic or supra-systemic pulmonary hypertension.
- Evaluation of pre- and post-saturations.
- Inhaled Nitric Oxide (iNO): consider only after echocardiogram and discussion with NICU Red Team and Cardiology. Contraindications include use in *isolated* LV dysfunction, LA enlargement, small left sided structures.
- Prostaglandins: Potential benefit exhibited in impaired right ventricular function and select congenital heart disease. Talk to NICU Red Team prior to starting.

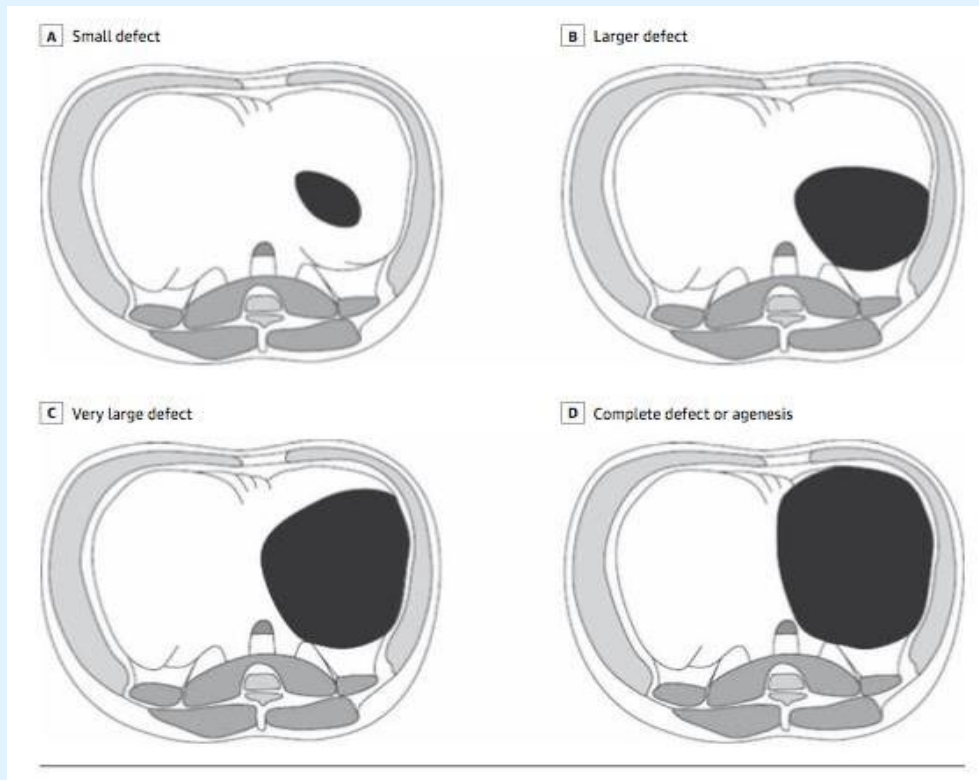
ECMO:

- Indications:
 - Inability to maintain pre-ductal saturations > 80% with acceptable ventilator settings.
 - Increased PaCO₂ and respiratory acidosis with pH < 7.15 despite efforts to optimize ventilator management.
 - Ventilator requirements of peak inspiratory pressure ≥ 26 cmH₂O or mean airway pressure ≥ 16 cmH₂O is required to achieve saturations > 80%.
 - Inadequate oxygen delivery with metabolic acidosis as evidenced by lactate > 3 mmol/L and pH < 7.15.
 - Systemic hypotension resistant to therapy.
- Inclusion criteria: Each patient will be evaluated on a case by case basis but general inclusion criteria are listed below:
 - Weight > 1.7 kg
 - Gestational age > 34 weeks
 - Absence of multiple or severe congenital anomalies or chromosomal anomalies.
 - Current head ultrasound with evidence of intraventricular hemorrhage ≤ Grade I.
 - If any of the above are not satisfied, infant may still be a candidate after multi-disciplinary discussion with pediatric surgery and ECMO team, as well as family counseling on risk/benefit ratio.

Perioperative – Goal: Early repair within 24 hours *only for those who are cannulated onto ECMO.
All other patients, timing of repair will be completion of transitional circulation and physiologically stable.

Surgical Repair:

- To be done at the bedside in the NICU in severely ill patient. Operating Room (OR) may be considered on an individual case basis in stable patients. Scheduling of cases early in the day allows for adequate support intra-operatively and post-operatively.
 - Patients exhibits evidence of physiologic stabilization, improvement of pulmonary hypertension.
 - Surgeons to document classification of defect size (A, B, C, D) in operative note (see below diagram).



Surgical Preparation:

- Preoperative labs completed within 24 hours prior to surgery and evaluated:
 - CBC with differential
 - BMP
 - Blood gas
 - Type & Cross (if not already completed)
 - If on ECMO, DIC profile required (PTT: 50 - 70 seconds; R-Time: 8 - 12 minutes)
- Pre-operative echocardiogram within 24 - 48 hours of surgical date.
- Order desired blood products to be in the OR or NICU at bedside during the procedure
 - Packed red blood cells, platelets, FFP (20 ml/kg of each)
- Clear space in patient room for anesthesia and surgery by removing nonessential equipment and staff.
- Ensure adequate IV access (2 PIV's or 1 PIV and 1 PICC) for administration of blood products and medications.
- Consider arterial access for monitoring of blood pressure.
- Replace TPN with D10½ Na⁺Cl⁻ to avoid electrolyte imbalances.
- Anesthesia to administer pre-operative antibiotics within 1 hour of incision.
- Make appropriate post-operative pain control plan and pre-order appropriate medications.

Post-Operative Care Management

- ❑ Remember to complete the VON QI OR Handoff Tool

Monitoring:

- Monitor post-operative vital signs per policy.
- Monitor for signs and symptoms of compartment syndrome: decrease distal pulses, abdominal distention, decreased urine output, skin discoloration.

Gastric Decompression:

- 10Fr salem sump orogastric or nasogastric tube to low continuous suction.
- NPO with salem sump until full return of bowel function.

Diagnostic Studies / Labs:

- CXR immediately post-operative.
- Blood gas immediately post-operative.
- CBC / BMP in the AM post-operative day #1.
 - Or earlier if clinically warranted

Oxygenation:

- Goal O₂ Saturations ≥ 90%.
- Continue pre and post-ductal saturation monitoring.

Ventilation:

- Target PaCO₂ 50 – 70 mmHg. Continue gentle ventilation.

Fluid Management:

- Continue pre-operative management of fluids and initiation of TPN after first post-operative void.

Hemodynamic Management:

- Closely monitor blood pressure.
- Consider fluid administration of vasopressor support as needed.

Antibiotics:

- Routine repair with no patch – Cefazolin for 24 hours; additional dose per surgeon discretion.
- Patch repair – Cefazolin for 24 hours post operative; additional doses per surgeon discretion.
- Open abdomen – consider Cefazolin until abdominal closure per surgeon discretion.
- Chest tube with patch – consider Cefazolin until discontinuation of chest tube per surgeon discretion.

Sedation: As clinically indicated – Refer to the NICU Pain Management Clinical Guideline

- Use of Acetaminophen IV 10mg/kg/dose q6hrs for 48 – 72 hrs postoperative pain control **AND** Start low dose continuous Morphine infusion 0.05 – 0.1 mg/kg/hr
- If infusions required pre-operative, utilize increased doses as needed to control post-operative pain.

Skin care:

- Notify surgery of any signs of erythema, drainage, bleeding or wound concerns.
- If sutures are place, contact surgery for removal plan/date.
- After sutures removed or surgical site has healed:
 - Apply Mepitel One (preemies) / Mepitac (post-term) to surgical sites once healed for scar therapy.
 - Change or re-apply after each bath.

Discharge Criteria

- Weight, length, head circumference updated
- Immunizations, hearing evaluation, EDAC screening completed
- Immunizations, hearing evaluations, Speech Therapy (ST) evaluation, CXR, ECHO, Head CT or MRI (if abnormal finding on HUS/seizures/abnormal neurologic findings/ECMO/patch repair)
- Follow up arranged for: RSV clinic, Cardiology, Pediatric Surgery and Outpatient ST

References

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