**Definition:** Congenital diaphragmatic hernia (CDH) is a relatively common malformation in which there is failure of complete fusion of the diaphragm during the prenatal period. This leads to protrusion of abdominal organs into the thoracic cavity which can result in lung hypoplasia and respiratory failure at birth. (1) CDH manifests on the left side in 75% of cases, which leads to possible herniation of the small and large bowel, the spleen, the stomach, the left lobe of the liver and, rarely, the kidney. Right-sided CDH can include herniation of the right lobe of the liver and possibly the bowel and/or kidney. (2) A bilateral defect is seen rarely. (3) Severity of disease depends upon the degree of abdominal organ herniation and lung hypoplasia.

**Epidemiology:** CDH occurs in approximately 1 in 3000 live births. Modern advances in technology and clinical practice for this condition has improved the reported overall survival to 79% (range: 69 – 93%), with isolated CDH having the highest survival rates. (4) In approximately 40% of cases, other associated anomalies are present in CDH patients, (3) including cardiac anomalies that are present in 25% of cases. (5) Musculoskeletal, neural tube, abdominal wall, craniofacial defects and urinary tract anomalies have been found in CDH patients.

**Etiology:** A variety of genetic abnormalities have been shown to cause isolated and non-isolated CDH. These abnormalities include large chromosome anomalies, microdeletions, microduplications, and mutations affecting single genes. CDH has also been shown to be a feature in a variety of genetic syndromes.

**Inclusion Criteria**
- Neonate diagnosed with CDH

**Exclusion Criteria**
- Infant not diagnosed with CDH

**Differential Diagnosis:** Chest x-ray results of CDH patients may be similar to that of a congenital cystic adenomatoid malformation (CCAM). The identification of abdominal organs in the thoracic cavity and a paucity of bowel in the abdomen solidifies diagnosis of CDH. (2)

**Diagnostic Evaluation:** The majority of CDH cases are diagnosed during antenatal ultrasound procedures during the second or third trimester. (6) Once the diagnosis is made, radiological ultrasound (US) and fetal magnetic resonance imaging (MRI) allows for determination of severity of disease by determining the percentage of herniated liver (%LH), observed-to-expected fetal lung volumes (O/E-TFLV) and the lung-head ratio (LHR). After birth, the diagnosis of CDH is made based on signs and symptoms with confirmation by x-ray. (2)

A detailed history and physical examination should be completed to assess the severity of the disease, to identify other congenital anomalies and to identify features that could suggest a specific genetic syndrome.

**History: Assess for**
- Relevant maternal history
- Results of antenatal imaging studies
- Results of genetic tests

**Physical Examination:** The following physical examination findings may be present in infants with congenital diaphragmatic hernia.
- Scaphoid abdomen
- Absence of breath sounds on the ipsilateral side
- Barrel-shaped chest
- Shifted cardiac sounds
- Bowel sounds in the chest

**Laboratory Tests**

<table>
<thead>
<tr>
<th>Lab</th>
<th>Delivery Room</th>
<th>Immediately upon NICU Admission</th>
<th>At 24 Hours of Life</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glucose</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Blood culture*</td>
<td>X</td>
<td></td>
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<tr>
<td>Arterial Blood Gas*</td>
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<tr>
<td>Serum Lactate#</td>
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<tr>
<td>CBC with Plat and Diff</td>
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<tr>
<td>Type and Screen</td>
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<td>X</td>
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<tr>
<td>BNP</td>
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<td>X</td>
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<tr>
<td>Chem 7</td>
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<td>X</td>
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<tr>
<td>Bilirubin Panel</td>
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<tr>
<td>Newborn Screen</td>
<td></td>
<td>X</td>
<td></td>
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<tr>
<td>Chromosomal Microarray Analysis</td>
<td></td>
<td>X</td>
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</tr>
</tbody>
</table>

*If concern for sepsis
#ABG & Lactate to be ordered at Q1 to Q4 (or greater) intervals depending on clinical status in first few hours of life
†BNP should be drawn within the first 24 hours of life and before surgery

**Ongoing Laboratory Tests**
- B-type natriuretic peptide (BNP) testing should be measured on day of life 1, postoperative day 1 and weekly thereafter. More frequent monitoring of BNP levels may be obtained as clinically indicated. (7-13)
In the neonatal period, cardiac cath may be considered upon
Depending on the gestational age at the time of CDH
Cardiac cath may be necessary if there is a need to perform
– Strong recommendation, moderate quality evidence
Cardiac cath should not be performed in a patient who
Practitioners should measure and document %LH, O/E-
A small percentage of the CDH population may require
It is recommended that a postnatal echocardiogram be
A cardiac cath may be considered, especially in the
STAT upon admission to NICU
The percentage of herniated liver (%LH) and observed-to-expected fetal lung volumes (O/E-TFLV) obtained from MRI should be used
A pre-established objective criteria should be used to score the severity of cardiac lesions. The presence of a major cardiac anomaly
Pulmonary venous abnormalities may be difficult to rule out
Practitioners should measure and document %LH, O/E-
TFLV, the side of the defect, total lung volume (TLV), lung-
to-head ratio (LHR), observed-to-expected lung-to-head ratio (O/E-LHR), stomach position and lung-to-thorax ratio for risk stratification, research, benchmarking and outcomes reporting in CDH patients. (14-37)
There should be a case-by-case determination of the management plan for CDH patients with comorbidities identified prenatally. This should be preferably discussed with the family prior to delivery and involve P-PACT if necessary. Limitations in care should be discussed in the antenatal visit.

Postnatal Echo
• It is recommended that a postnatal echocardiogram be performed within the first 24-48 hours after birth. It may be obtained earlier if the neonate exhibits a sub-optimal response to the usual therapy, especially if a prenatal echo was suboptimal or not performed. (38-42)
• A postnatal echo may be obtained electively, at a later, convenient time, if the neonate does not exhibit undue difficulties in ventilation and oxygenation, especially if a good quality fetal echo was performed.

Critical Points of Evidence

Evidence Supports

Prenatal Testing
• The percentage of herniated liver (%LH) and observed-to-expected fetal lung volumes (O/E-TFLV) obtained from MRI should be used to stratify CDH severity which can be used in prenatal counseling to educate families on disease severity and mortality risks in CDH patients. (14-26) – Strong recommendation, moderate quality evidence
• Practitioners should measure and document the %LH, O/E-TFLV, side of the defect, total lung volume (TLV), lung-to-head ratio (LHR), observed-to-expected lung-to-head ratio (O/E-LHR), stomach position and lung-to-thorax ratio for risk stratification, research, benchmarking and outcomes reporting in CDH patients. (14-39) – Strong recommendation, moderate quality evidence
• A pre-established objective criteria should be used to score the severity of cardiac lesions. The presence of a major cardiac anomaly is associated with an increased risk of mortality. (32,43-46) – Strong recommendation, low quality evidence
• The oxygenation index should continue to be used for management decisions in CDH patients. (46-49) – Strong recommendation, low quality evidence
• The Brindle Prediction Score (see appendix) should be used to educate families on the categorical (low, intermediate, or high) mortality risk in CDH patients. (14,50) – Strong recommendation, low quality evidence

Initial Management

• Clinicians may consider the use of surfactant in CDH patients with a history of Fetal Endotracheal Occlusion (FETO) or preterm birth (<37 weeks gestation). (5,51-57) – Weak recommendation, low quality evidence
  o Remarks: The fetus that undergoes FETO can have a decrease in type II pneumocytes which can cause surfactant deficiency. Patients receiving this procedure during the prenatal period may benefit from postnatal surfactant administration. Response to therapy guides dosing of surfactant. Unless there were negative consequences to initial administration, clinicians should consider a second dose. If the patient does not exhibit a positive response to the first two doses of surfactant, do not administer additional doses. (58,59)

Laboratory Testing and Imaging

• B-type natriuretic peptide (BNP) testing should be measured on day of life 1, postoperative day 1 and weekly thereafter. More frequent monitoring of BNP levels may be obtained as clinically indicated. (7-13) – Strong recommendation, very low quality evidence
• Echocardiogram should be performed within 24 to 48 hours of life, one week after CDH repair, and within four weeks of discharge. Additional follow-up echos should be obtained every 1 to 2 weeks for patients with severe pulmonary hypertension until there is evidence of resolution with pulmonary pressures <1/2 systemic. (18-40) – Strong recommendation, very low quality evidence

Systemic Hypotension Management

• Titrate dopamine to manage systemic hypotension in CDH patients during the acute phase. (55,60-67) – Strong recommendation, low quality evidence

ECMO

• Administer a one-time dose of cefazolin within one hour of cannulation for ECMO in CDH patients. Do not routinely use continued prophylactic antibiotics for patients on ECMO support. (68-70) – Strong recommendation, very low quality evidence

Surgical Repair

• In non-repaired CDH patients on ECMO, surgical repair of the diaphragm should be completed within 48 hours of ECMO cannulation. (1,5,55,63,64,66,67,71-78) – Strong recommendation, low quality evidence
• In CDH patients not requiring ECMO, surgical repair of the diaphragm should be performed when the patient is physiologically stable defined as mean blood pressure normal for gestational age, preductal oxygen saturations between 85-95% on FiO2 below 50%, lactate below 3 mmol/L, urine output greater than 2 mL/kg and less than a 10% gradient between pre- and post-ductal saturations. (1,5,55,63,64,66,67,71-78) – Strong recommendation, low quality evidence
• In CDH patients that are not able to achieve a tension-free primary closure of the diaphragm, a patch repair should be completed with an appropriately sized domed patch. (76-79) – Strong recommendation, low quality evidence
• CDH patients with large defects should be repaired using an open surgical technique. (78,82-85) – Strong recommendation, moderate quality evidence
• The use of minimally invasive surgery (MIS) may be considered in patients that meet the following criteria: mean blood pressure normal for gestational age without use of inotropes, pre-ductal oxygen saturations between 85-95% on FiO2 below 40% without use of inhaled nitric oxide, lactate below 3 mmol/L, urine output greater than 2 mL/kg/hr, and less than a 5% gradient between pre- and post- ductal saturations. (1,5,55,63,64,66,67,71-78) – Weak recommendation, moderate quality evidence

Evidence Lacking/Inconclusive

Management Plan

• Determine management plan for severe cardiac lesions first before proceeding to establish plan of care for congenital diaphragmatic hernia. – Consensus recommendation

Ventilation Strategy

• Patients with CDH should be initially ventilated with the conventional ventilator using the AC/VG mode with initial settings of PEEP 5-6 cmH2O, TV 4-5 mL/kg, back-up rate 40 breaths/min, IT 0.3 seconds, and FiO2 adjusted for target preductal saturations of ≥80%. (5,55,63,67,78,87-91) – Strong recommendation, low quality evidence
• Tidal volume should be adjusted to meet optimal physiological monitoring parameters. (5,55,63,67,78,87-91) – Strong recommendation, low quality evidence
• Clinicians should consider switching the mode of ventilation to HFOV for CDH patients that cannot achieve target PCO2 on conventional ventilation with PIP ≤28. (5,55,63,67,78,87-91) – Weak recommendation, low quality evidence
  o Once on HFOV, increase MAP (to a max MAP of 17) and DeltaP as required to achieve physiological monitoring parameters.
• Clinicians should consider ECMO for CDH patients that cannot achieve saturations and/or blood gas targets with maximal HFOV support. (5,55,63,67,78,87-91) – Weak recommendation, low quality evidence
• Patients with CDH should have the following targets for physiological monitoring parameters: pre-ductal oxygen saturations ≥80% for the first two hours of life. Thereafter, pre-ductal saturations should be ≥85%; pH >7.20; pCO2 50 to 70; and pO2 40 to 90. (5,55,63,67,78,87-91) – Strong recommendation, very low quality evidence
• Patients with CDH should be initially ventilated with 100% FiO2. – Consensus recommendation
• Pre-ductal saturations should be targeted at ≥70% for the first ten minutes after birth; increasing to ≥80% for the first two hours of life. Thereafter, pre-ductal oxygen saturations should be ≥85%. – Consensus recommendation

Venous Access and Fluid Management
• Peripheral venous access should be established in the delivery room for CDH patients. – Consensus recommendation
• Umbilical lines and elective/non-emergent procedures should be completed in the NICU. – Consensus recommendation
• An umbilical venous catheter (UVC) and an appropriately placed umbilical arterial catheter (UAC) should be initially inserted in CDH patients with a need for central venous access. If correct UVC position cannot be achieved, a temporary low-lying UVC can be initially utilized until a sufficient alternative is available. (102-103) – Strong recommendation, low quality evidence
• A peripherally inserted central catheter (PICC) and an arterial line should be placed by the NICU Vascular Access Team (VAT) in CDH patients with a need for long-term central venous access once stabilization occurs. – Consensus recommendation
• Initial fluid intake for CDH patients should be 65 mL/kg/day. – Consensus recommendation

Pulmonary Hypertension Management
• In infants with congenital diaphragmatic hernia with an oxygenation index (OI) >25, preductal saturations <90% on 100% FiO₂ or a gradient between pre- and postductal saturations ≥10%, consider the use of inhaled nitric oxide to improve oxygenation. (5,38,55,63-67,78,110,112-114) – Weak recommendation, low quality evidence
• In infants with congenital diaphragmatic hernia, consider the use of sildenafil for continued clinically significant pulmonary hypertension beyond the acute phase as evidenced by a preductal/postductal saturation difference of >10 points, in those infants who fail weaning from inhaled nitric oxide or to facilitate weaning from nitric oxide. (5,38,55,63-67,78,110,112-114) – Weak recommendation, low quality evidence
• In infants with congenital diaphragmatic hernia requiring VA ECMO, inhaled nitric oxide should be discontinued once VA ECMO support is initiated and restarted when recruiting the lungs in preparation for discontinuing ECMO. (55,112-114) – Strong recommendation, very low quality evidence
• In patients on VV ECMO, consider the use of inhaled nitric oxide throughout ECMO run. (5,112-114) – Weak recommendation, very low quality evidence
• Remarks: Response to inhaled nitric oxide should be assessed in all users within one hour of initiation. Responders will exhibit a >5% increase in preductal saturations, an increase in PaO₂ by 10 torr (obtained from the same pre- or post-ductal source) or a decrease in pre/post ductal saturation gradient to <10%. Inhaled nitric oxide should be weaned based upon patient classification as responder or non-responder.

Systemic Hypotension Management
• Consider administration of hydrocortisone (1mg/kg IV every 8 hours) in CDH patients on a dose of greater than or equal to 10 mcg/kg/min of dopamine with continued systemic hypotension. (5,63-67,115-117) – Weak recommendation, very low quality evidence
• Consider the use of epinephrine in CDH patients on maximum dopamine dose and hydrocortisone. (55,63-67,118) – Weak recommendation, very low quality evidence
• Remarks: If the patient has vasopressor refractory hypotension, an echo should be obtained and the treated according to findings.

Evidence Against

Prenatal Testing
• The Brindle Prediction Score (see appendix) should not be used for individual patient decisions regarding the use of ECMO or other medical management. (14,90) – Strong recommendation, low quality evidence
• The SNAP II Score, Wilford Hall/Santa Rosa Clinical Prediction Formula, Congenital Diaphragmatic Hernia Study Group (CDHSG) formula, the best PaCO₂ at 1 hour of life and 24 hours of life, and the highest preductal Q₂ saturation value during the first 24 hours of life should not be used to predict survival or make decisions about treatment options for CDH patients. (46,119-130) – Strong recommendation, low quality evidence

Initial Management
• Surfactant should not be routinely used in the non-FETO term CDH patient at birth. (5,51-57) – Strong recommendation, low quality evidence
• Information obtained from near infrared spectroscopy (NIRS) readings should not be used for patient care management. (131) – Strong recommendation, low quality evidence

Pulmonary Hypertension Management
• Prostaglandin E (PGE) should not be routinely used in the acute phase of CDH treatment. (5,38,55,63-67,78,132,133) – Strong recommendation, very low quality evidence
• Epoprostenol should not be routinely used for the treatment of pulmonary hypertension in neonates with congenital diaphragmatic hernia. (5,38,55,63-67,78,134-136) – Strong recommendation with very low quality evidence

Imaging
• Cardiac cath is not recommended in the acutely presenting CDH neonate in whom congenital heart disease has been ruled out by echocardiography. (38-42) – Strong recommendation, very low quality evidence

Ventilation Strategy
• Heliox should not be routinely used in CDH patients. (137-141) – Strong recommendation, low quality evidence
**Condition-Specific Elements of Clinical Management**

**Initial Management in the Delivery Room:**
- Intubate immediately and ventilate with conventional ventilator on AC/VG mode on 100% FiO2 (1,5,55,63,64,66,67,71-78)
  - PEEP: 5-6 cmH2O
  - TV: 4-5 cc/kg
  - MAX PIP: 28
  - Back-up rate: 40 breaths/minute
  - IT: 0.3 seconds
  - FiO2 - adjust for target pre-ductal saturations of ≥80%
- Insert 10 french (FR) repogle and place to low-intermittent suction.
- Establish peripheral venous access and begin 10% dextrose solution. Total initial total fluid intake should be 65 mL/kg/day.
- Continually assess for respiratory distress or low pre-ductal saturations. Pre-ductal saturations should be targeted at ≥70% for the first ten minutes after birth; increasing to ≥80% for the first two hours of life.
- Adjust TV to improve distress and increase pre-ductal saturations to optimal targets
- If continued distress, adjust TV and PEEP.
- Surfactant should not be routinely used. If history of FETO or <37 weeks gestation, consider surfactant administration.5,51,57
- If distress continues, adjust ventilator and/or hand bagging as needed. Plan for HFOV following admission to WT NICU.
- Umbilical lines and elective procedures should be completed in the NICU.
- Goal for timespan of initial resuscitation in L&D is 30 minutes or less.

**Acute Phase (Birth to Surgical Repair):**

**General**
- Admit to NICU 4
- 10 FR repogle to low-intermittent suction (LIS) if not already done
- Insert appropriately placed UVC and UAC. If umbilical lines not appropriately placed, obtain a PICC line and/or PAL.102,103
- Consult ECMO surgeon, Neo ECMO clinician, ECLS Primer and Pulmonary Hypertension team.
- Monitor pre- and post-ductal saturations
- Administer erythromycin and Vitamin K

**Ventilation Management** 5,55,63-78,87-81
- Tidal volume should be adjusted to meet optimal physiologic parameters of:
  - pre-ductal saturations ≥85% after two hours of life
  - pH >7.20
  - pCO2 between 50-70
  - pO2 between 40-90
- Consider switching mode of ventilation to HFOV if patient cannot achieve optimal physiologic parameters on PIP ≤28.
- Initial HFOV settings are:
  - MAP 13 (or 2 above that on conventional ventilator)
  - IT 0.3
  - Hz 10
  - DeltaP sufficient to produce perceptible “jiggle” to upper abdominal area.
- Once on HFOV, increase MAP (to a max MAP of 17) and DeltaP as required to achieve physiological monitoring parameters.

**Treatment for Pulmonary Hypertension**
- Consider the use of inhaled nitric oxide to improve oxygenation if oxygenation index (OI) >25, pre-ductal saturations <90% on 100% FiO2 or a gradient between pre- and post-ductal saturations ≥10%. (5,38,55,63-78,104-109)
- Response to inhaled nitric oxide should be assessed in all users. Responders will exhibit a >5% increase in preductal saturations, an increase in PaO2 by 10 torr or a decrease in pre/post ductal saturation gradient to <10%. Inhaled nitric oxide should be weaned based upon patient classification as responder or non-responder.

**ECMO**
- Indications for ECMO include oxygenation index (OI) >40 on two separate measurements, PO2 persistently <40 mmHg or lactate rising above 3.
- Administer a one-time dose of cefazolin within one hour of administration.
- Consider switching mode of ventilation to HFOV if patient is not a responder or non-responder.
- Most CDH patients should have mean BP normal for gestational age.
- Titrare continuous dopamine to achieve optimal blood pressure. (55,80-87)
- Once dopamine reaches 10 mcg/kg/min., consider administration of hydrocortisone. (57,69-71,117)
- Dopamine can continue to be titrated until a MAX of 20 mcg/kg/min to obtain optimal blood pressure.
- If optimal blood pressure is not obtained on MAX dopamine and hydrocortisone, consider starting continuous epinephrine. (55,63-67,118)

**Surgical Repair**
- Surgical repair of the diaphragm should be performed when the patient is physiologically stable defined as below. (1,5,55,63,64,66,67,71-78)
  - Mean blood pressure normal for gestational age
  - Pre-ductal oxygen saturations between 85-95% on FiO2 below 50%
  - Lactate below 3 mmol/L
  - Urine output greater than 2 mL/kg/hr
  - Less than a 10% gradient between pre- and post-ductal saturations
- Patients who meet the additional requirements of pre-ductal oxygen saturations between 85-96% on FiO2 40% without the use of nitric oxide, mean BP normal for age without use of inotropes and less than a 5% gradient between pre- and post-ductal saturations may also be considered for minimally invasive surgery. (1,5,55,63,64,66,71-78)
- In non-repaired CDH patients on ECMO, surgical repair of the diaphragm should be completed less than 48 hours after ECMO cannulation. (1,5,55,63,64,66,71-78)
- CDH patients with large defects should be repaired using an open surgical technique. (78,82-86)
- A patch repair with an appropriately sized domed patch should be undertaken if a tension-free primary closure is not achievable during surgery. (78,81)

**Post-Acute / Chronic Phase:**

**Treatment for Pulmonary Hypertension**
- Consider the use of sildenafil for continued clinically significant pulmonary hypertension beyond the acute phase
as evidenced by a preductal/postductal saturation difference of >10 points or in those infants who fail weaning from inhaled nitric oxide. (5,38,55,63-67,78,110,111)

- Consider the use of bosentan as third line treatment for refractory pulmonary hypertension in CDH patients without liver dysfunction and with the ability to receive enteral medications. (142)

- In infants with congenital diaphragmatic hernia requiring VA ECMO, inhaled nitric oxide should be discontinued after repair when the patient is on ventilator rest settings and restarted when recruiting the lungs in preparation for discontinuing ECMO. Inhaled nitric oxide should be weaned based upon patient classification as responder or non-responder. (55,112-114)

**Discharge Criteria**

Discharge planning should begin upon admission and continue throughout hospital stay. The decision to discharge should be discussed with the pulmonary and surgical teams. Coordination of discharge is essential as some treatments may take as long as four weeks for approval from payers.

- Stable cardio/respiratory for at least two weeks that can be safely delivered in the home environment.
  - FiO₂ ≤40% with home ventilator
  - NC 1 liter per minute (LPM) or less
- Stable nutrition program with documented weight gain and growth velocity.
- Appropriate training and documented education of family or guardian.
- Prescriptions should be filled and in the possession of parent or guardian at the time of discharge.
- Designated primary care physician and follow-up plan with appropriate teams.
- Consider rooming-in for patients with complicated home care

**Consults/Referrals**

- Surgery
- Pulmonary
- Cardiology
- Pulmonary Hypertension Team
- Genetics, as indicated

**Follow-Up Care:**

- Primary Care Physician
- Surgery
- Pulmonary Hypertension Team
- Developmental

**Measures:**

**Process**

- Time of birth to admit to NICU
- Rate of antenatal consults by neonatology or relevant service (Cardiology, Surgery)
- Rate of surfactant use in the first 72 hours of life
- Timing of first echo

**Outcome**

- Mortality rate
- Mean and median duration of ventilation
- ECMO rate
- Length of stay
- Duration of ECMO
- ECMO complications
- Rate of discharge on pulmonary hypertension treatment
- Rate of discharge on enteral feeding
Medication Dosing

**Surfactant**

Surfactant should not be routinely used in the non-FETO term CDH patient at birth. Clinicians may consider surfactant in CDH patients with a history of Fetal Endotracheal Occlusion (FETO) or preterm birth (<37 weeks gestation).

<table>
<thead>
<tr>
<th>Medication</th>
<th>Route</th>
<th>Dosing Information</th>
</tr>
</thead>
<tbody>
<tr>
<td>Curosurf Inhalation</td>
<td>Inhalation</td>
<td>2.5 mL/kg via endotracheal tube for the first dose. Decrease dose to 1.25 mL/kg for subsequent doses. Response to therapy guides dosing of surfactant.</td>
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</tbody>
</table>

**Systemic Hypotension Treatment**

<table>
<thead>
<tr>
<th>Medication</th>
<th>Route</th>
<th>Dosing Information</th>
</tr>
</thead>
<tbody>
<tr>
<td>DOPamine</td>
<td>Continuous IV infusion</td>
<td>5 mcg/kg/min. Titrate to MAX of 20 mcg/kg/min.</td>
</tr>
<tr>
<td>Hydrocortisone</td>
<td>IV</td>
<td>1 mg/kg every 8 hours</td>
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<tr>
<td>Note: Consider administration of hydrocortisone once dopamine is titrated to 10 mcg/kg/min or greater.</td>
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</tr>
<tr>
<td>EPINEPHrine</td>
<td>Continuous IV infusion</td>
<td>0.05 mcg/kg/min. Titrate to MAX of 1 mcg/kg/min.</td>
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</tbody>
</table>

**Pulmonary Hypertension Treatment – Acute Phase**

<table>
<thead>
<tr>
<th>Medication</th>
<th>Route</th>
<th>Dosing Information</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nitric Oxide</td>
<td>Inhalation</td>
<td>Start with initial dose of 20 ppm; titrate down to lowest effective dose</td>
</tr>
</tbody>
</table>

**ECMO Preparation**

<table>
<thead>
<tr>
<th>Medication</th>
<th>Route</th>
<th>Dosing Information</th>
</tr>
</thead>
<tbody>
<tr>
<td>CeFAZolin</td>
<td>IV</td>
<td>25-30 mg/kg IV ONCE 30-60 minutes before procedure</td>
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</table>
TCH Evidence-Based Outcomes Center
Clinical Algorithm for Initial Management of Congenital Diaphragmatic Hernia Patients

**Labor and Delivery Algorithm**

- **CDH patient delivered at TCH LD unit**
  - Place on giraffe warmer
  - Intubate immediately
  - Ventilate with conventional ventilator on VG/AC mode on 100% FiO2*
  - Insert 10 FR repogle and place to intermittent suction

- **CDH Initial Ventilator Settings**
  - Conventional ventilator AC/VG
  - PEEP: 5 – 6 cmH2O
  - TV: 4 – 5 cc/kg
  - MAX PP: 28
  - Back-up rate: 40 breaths/minute
  - IT: 0.3 seconds
  - FiO2 – adjust for target pre-ductal saturations of ≥80%

- **Start D10W IV at 65mL/kg/day**
  - Transfer pt to West Tower NICU 4

- **Resp distress or pre ductal saturations <80% ≠**
  - Establish PIV access
  - Adjust TV and PEEP per CDH ventilator guidelines to achieve targets
  - Surfactant should not be routinely administered to non-FETO term CDH patients^*
  - If history of FETO or <37 weeks gestation, consider surfactant administration^*

- **Continued distress or saturations <80%**
  - **Yes**
    - Surfactant should not be routinely administered to non-FETO term CDH patients^*
    - If history of FETO or <37 weeks gestation, consider surfactant administration^*
  - **No**
    - Adjust TV and PEEP per CDH ventilator guidelines to achieve targets
    - Plan for HFOV following admission to WT NICU

- **Continued distress or saturations <80% ≠**
  - **Yes**
    - Adjust ventilator and/or hand bagging as needed
    - Plan for HFOV following admission to WT NICU
  - **No**
    - Establish IV access

**Goal for timespan of initial resuscitation in L&D is 30 minutes or less**

- **Yes**
  - Preductal oxygen saturations should be targeted for ≥70% for the first ten minutes after birth and thereafter >80% for the first two hours of life*
  - Patient may be transferred to the NICU without IV access if unable to obtain after four attempts
  - West Tower NICU 4 should be notified ahead of transport to prepare for line placement once patient arrives in unit

- **No**
  - Plan for HFOV following admission to WT NICU

**5 Min of Life**

**10 Min of Life**

**20 Min of Life**
Clinical Algorithm for the Acute Management of Congenital Diaphragmatic Hernia Patients Prior to ECMO

Inpatient Algorithm

Admit to NICU 4
Initiate conventional mechanical ventilation
Adjust tidal volume to achieve physiologic monitoring parameter goals
Place 10 French repogle to LIS if not done
Insert UVC and appropriately placed UAC if not already done
Consult ECMO surgeon, Neo ECMO clinician and ECLS specialist
Monitor pre and postdural saturations
Administer erythromycin and Vitamin K

Obtain a PICC and/or PAL if umbilical lines not appropriately placed
Draw ABG, lactate and NICU admission labs† once umbilical lines placed
Obtain CXR and KUB
Obtain HUS in in first 24 hours of life
If at risk for sepsis, obtain a blood culture and administer ampicillin and gentamicin
Obtain an echo within 24 to 48 hours after birth

Initiate HFOV per CDH ventilation guidelines
Consult ECMO Clinician if not already done
ABG in 30 minutes
CXR in one hour

Hypotension?

Yes

Titrated dopamine to achieve optimal blood pressure
Once dopamine reaches 20 mcg/kg/min, start hydrocortisone

No

Continue to monitor

Does patient meet criteria for repair?

Yes

Repair complete

No

Continuously assess for readiness for repair
When criteria for surgery met proceed to next step

Resp distress OR physiologic parameters out of range?

No

Adjust TV and PEEP per CDH Ventilation guidelines

Yes

Criteria to start iNO met?

Yes

Start iNO at 20 ppm
ABG within one hour to evaluate response to NO

No

Continue to titrate dopamine
Once dopamine reaches 20 mcg/kg/min, consider starting epinephrine
If hypotension unresolved with epinephrine, obtain an echo and treat accordingly

Hypotension continues?

Yes

No

Continue to monitor

Criteria for MIS:

Mean BP normal for gestational age without inotropes
Pre-ductal sats 85-95% on FiO2 < 40% without iNO
Lactate < 3
Urine output > 2 ml/kg/hr
Less than 5% gradient in pre/post-ductal sats

Criteria to start iNO met?

Yes

Continue

No

Does patient meet criteria for repair?

Yes

No

Surgeon to determine eligibility for MIS or open repair

Repair complete

Criteria for Repair:

Mean BP normal for gestational age
Pre-ductal sats 85-95% on FiO2 < 50%
Lactate < 3
Urine output > 2 ml/kg/hr
Less than 30% gradient in pre/post-ductal sats

Criteria for MIS:

Mean BP normal for gestational age without inotropes
Pre-ductal sats 85-95% on FiO2 < 40% without iNO
Lactate < 3
Urine output > 2 ml/kg/hr
Less than 5% gradient in pre/post-ductal sats

Optimal Physiologic Monitoring Parameters

Preductal sats ≥85%
pH >7.20
pCO2: 50 to 70
pO2: 40 to 90

HFOV Settings

MAP 13 (or 2 above that on conventional ventilator)
MAX MAP: 17
IT: 0.3
Delta P: Sufficient to produce perceptible “jiggle” to upper abdominal area

Additional ECMO Criterial

Start ECMO; administer one dose of cefazolin within one hour of cannulation
ABG STAT
CRRT STAT

Criteria for iNO – any item below

OI > 25
Preductal sats < 90% on 100% FiO2
Pre/post dural gradient > 50%

Legend

NICU – neonatal intensive care unit
BUN – blood urea nitrogen
Cr – creatinine
Hct – hematocrit
Hb – hemoglobin
Hct – hematocrit
IOP – intraocular pressure
LVEF – left ventricular ejection fraction
MIS – minimally invasive surgery
PICC – peripherally inserted central catheter
AC/VG – assist control volume guarantee
OI – oxygenation index

Continued

Resp distress OR physiologic parameters out of range?

MIS – minimally invasive surgery
PICC – peripherally inserted central catheter
AC/VG – assist control volume guarantee
OI – oxygenation index

Additional ECMO Criterial

Start ECMO; administer one dose of cefazolin within one hour of cannulation
ABG STAT
CRRT STAT

Patient placed on ECMO

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References


DATE: December 2017

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64. UT Health Science Center at Houston. (2015). Management of infants with congenital diaphragmatic hernia from birth to surgery.


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DATE: December 2017


Clinical Standards Preparation

This clinical standard was prepared by the Evidence-Based Outcomes Center (EBOC) team in collaboration with content experts at Texas Children’s Hospital. Development of this clinical standard supports the TCH Quality and Patient Safety Program initiative to promote clinical standards and outcomes that build a culture of quality and safety within the organization.

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Development Process

This clinical standard was developed using the process outlined in the EBOC Manual. The literature appraisal documents the following steps:

1. Review Preparation
   - PICO questions established
   - Evidence search confirmed with content experts

2. Review of Existing Internal and External Guidelines
   - Guidelines for Acute Care of the Neonate, Baylor College of Medicine Section of Neonatology; Management of CDH Infants, TCH Baylor Physicians; Standardized Postnatal Management of Infants with Congenital Diaphragmatic Hernia in Europe, CDH Euro-Consortium; Surfactant Replacement Therapy for Preterm and Term Neonates with Respiratory Distress, American Academy of Pediatrics; Surfactant Replacement Therapy, American Association of Respiratory Care; NICU Clinical Guidelines Respiratory Problems and Management Congenital Diaphragmatic Hernia, King Edward Memorial Hospital; Management of Congenital Diaphragmatic Hernia: A Systematic Review from the APSA Outcomes and Evidence Based Practice Committee, American Pediatric Surgical Association; CDH Protocol, Cincinnati Children’s Hospital; Management of Infants with Congenital Diaphragmatic Hernia from Birth to Surgery, UT Health Science Center at Houston; Congenital Diaphragmatic Hernia Management, Children’s Hospital Network; Pediatric Pulmonary Hypertension, American Heart Association and American Thoracic Society; Expert Consensus Statement on the Diagnosis and Treatment of Pediatric Pulmonary Hypertension, The European Paediatric Pulmonary Vascular Disease Network; Pulmonary Hypertension Associated with Acute or Chronic Lung Diseases in Preterm or Term Neonate and Infant, The European Paediatric Pulmonary Vascular Disease Network; General Guidelines for all ECLS Cases, Extracorporeal Life Support Organization; Infection Control and Extracorporeal Life Support, Extracorporeal Life Support Organization; Thoracoscopic Repair of Congenital Diaphragmatic Hernia in Neonates, National Institute for Health and Clinical Excellence

3. Literature Review of Relevant Evidence
   - Searched: PubMed, Cochrane Library, Google Scholar, Cinahl, Guideline Clearing House

4. Critically Analyze the Evidence
   - 14 meta-analyses, 4 randomized controlled trials, and 103 nonrandomized studies

5. Summarize the Evidence
   - Materials used in the development of the guideline, evidence summary, and order sets are maintained in a congenital diaphragmatic hernia evidence-based review manual within EBOC.

Evaluating the Quality of the Evidence

Published clinical guidelines were evaluated for this review using the AGREE II criteria. The summary of these guidelines are included in the literature appraisal. AGREE II criteria evaluate Guideline Scope and Purpose, Stakeholder Involvement, Rigor of Development, Clarity and Presentation, Applicability, and Editorial Independence using a 4-point Likert scale. The higher the score, the more comprehensive the guideline.

This clinical standard specifically summarizes the evidence in support of or against specific interventions and identifies where evidence is lacking/inconclusive. The following categories describe how research findings provide support for treatment interventions.

“Evidence Supports” provides clear evidence that the benefits of the intervention exceed harm.

“Evidence Against” provides clear evidence that the intervention is likely to be ineffective or that it is harmful.

“Evidence Lacking/Inconclusive” indicates there is currently insufficient data or inadequate data to support or refute a specific intervention.

The GRADE criteria were utilized to evaluate the body of evidence used to make practice recommendations. The table below defines how the quality of the evidence is rated and how a strong versus weak recommendation is established. The literature appraisal reflects the critical points of evidence.
### Recommendation

<table>
<thead>
<tr>
<th>Quality</th>
<th>Type of Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>STRONG</td>
<td>Desirable effects clearly outweigh undesirable effects or vice versa</td>
</tr>
<tr>
<td>WEAK</td>
<td>Desirable effects closely balanced with undesirable effects</td>
</tr>
<tr>
<td>High</td>
<td>Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies</td>
</tr>
<tr>
<td>Moderate</td>
<td>Evidence from RCTs with important limitations (e.g., inconsistent results, methodological flaws, indirect evidence, or imprecise results) or unusually strong evidence from unbiased observational studies</td>
</tr>
<tr>
<td>Low</td>
<td>Evidence for at least 1 critical outcome from observational studies, RCTs with serious flaws or indirect evidence</td>
</tr>
<tr>
<td>Very Low</td>
<td>Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence</td>
</tr>
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</table>

### Recommendations

Practice recommendations were directed by the existing evidence and consensus amongst the content experts. Patient and family preferences were included when possible. The Content Expert Team and EBOC team remain aware of the controversies in the diagnosis/management of congenital diaphragmatic hernia in children. When evidence is lacking, options in care are provided in the clinical standard and the accompanying order sets (if applicable).

### Approval Process

Clinical standards are reviewed and approved by hospital committees as deemed appropriate for its intended use. Clinical standards are reviewed as necessary within EBOC at Texas Children’s Hospital. Content Expert Teams are involved with every review and update.

### Disclaimer

Practice recommendations are based upon the evidence available at the time the guideline was developed. Clinical standards (guidelines, summaries, or pathways) do not set out the standard of care, and are not intended to be used to dictate a course of care. Each physician/practitioner must use his or her independent judgment in the management of any specific patient and is responsible, in consultation with the patient and/or the patient family, to make the ultimate judgment regarding care.

### Version History

<table>
<thead>
<tr>
<th>Date</th>
<th>Action</th>
<th>Comments</th>
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<tbody>
<tr>
<td>June 2016</td>
<td>Evidence summary completed</td>
<td>Content related to initial management of CDH patients</td>
</tr>
<tr>
<td>Dec 2017</td>
<td>Guideline completed</td>
<td>Added acute management until surgical repair content</td>
</tr>
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</table>
Appendix

The observed-to-expected total lung volume (O/E TFLV) and percent liver herniation (%LH) has been used for prenatal risk stratification in patients with congenital diaphragmatic hernia (29). Below are the risk stratification categories utilized at Texas Children’s Hospital.

<table>
<thead>
<tr>
<th>Severity</th>
<th>O/E TFLV</th>
<th>LH%</th>
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<tbody>
<tr>
<td>Mild</td>
<td>&gt;32%</td>
<td>&lt;21%</td>
</tr>
<tr>
<td>Moderate</td>
<td>≥32%</td>
<td>&gt;21%</td>
</tr>
<tr>
<td>Moderate</td>
<td>&lt;32%</td>
<td>≤21%</td>
</tr>
<tr>
<td>Severe</td>
<td>&lt;32%</td>
<td>&gt;21%</td>
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</table>