Management of Congenital Diaphragmatic Hernia – Neonatal – Inpatient - Consensus Care Guideline

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Content Experts:
Jamie Limjoco, MD- Pediatrics- Neonatal ICU
Email address: jjlimjoco@pediatrics.wisc.edu

C. Lydia Wraight, MD- Pediatrics- Neonatal ICU
Email address: wraight@pediatrics.wisc.edu

Charles Leys, MD- Surgery- Pediatric Surgery
Email address: leys@surgery.wisc.edu

Workgroup Members:
Michael Wilhelm, MD - Pediatrics- ICU
Charles Bergstrom, MD - Pediatrics-PICU
Ryan McAdams, MD - Neonatology
Inna Lobeck, MD – Pediatric Surgery
Hau Le, MD - Pediatric Surgery

Reviewers:
Petros Anagnostopoulos, MD - Cardiothoracic Surgery
Shardha Srinivasan, MD – Pediatric Cardiology
Peter Nichol, MD - Pediatric Surgery
Sushant Srinivasan, MD - PICU
Laura Bodine, MS, RD, CNSC, CD- Clinical Nutrition- AFCH
Emily Russart, RN – Neonatal ICU
Tim Elgin, MD – Neonatal ICU
Ben Walker, MD- Anesthesiology
Josh Vanderloo, PharmD - Drug Policy Program
Suzanne Hoffman, MT, ASCP- Clinical Labs - Hem/Special Coagulation
Laura Konkol, MSN, RN- Nursing - Neonatal ICU
Igor Iruretagoyena, MD – OB-GYN - Perinatal

Contact for Changes:
Center for Clinical Knowledge Management (CCKM)
Email Address: CCKM@uwhealth.org

Guideline Author:
Lee Skrupky, PharmD – CCKM

Committee Approvals/Dates:
Clinical Knowledge Management (CKM) Council (5/25/2023)
NICU Clinical Guidelines and Quality Improvement Committee 4/5/2023

Introduction

Congenital diaphragmatic hernia (CDH) is a congenital condition that affects 1 in every 2,000-3,000 infants each year.[1] Patients are born with a hole in their diaphragm that allows the abdominal organs to move into the chest, causing pulmonary hypoplasia. Approximately 50-70% of CDH cases are "isolated", in which the hernia is the only defect; in the remainder of cases there are additional complications including significant structural and/or genetic abnormalities.[2,3] This condition requires surgical repair of the diaphragm and the detailed coordination of care between neonatology, intensive care, and surgery.

Scope

Intended Users: Physicians, Advanced Practice Providers, Nursing, Pharmacists, Respiratory Therapists

Objectives: To outline evidence-based recommendations and reduce practice variation in the management of patients with congenital diaphragmatic hernia across UW Health

Target Population: Neonatal patients with congenital diaphragmatic hernia.

Clinical Questions Considered:

- What is included as a part of prenatal assessment and planning?
- What are key management considerations for infants with CDH immediately following delivery?
- What approaches to ventilator management (and related care measures) are recommended throughout the phases of care for infants with CDH?
- What treatment considerations are recommended to manage pulmonary hypertension and hemodynamic alterations in babies with CDH?
- When should extracorporeal membrane oxygenation (ECMO) be considered?
- What are the key surgical considerations for repair of CDH infants?
- What follow-up is recommended following discharge of babies with CDH?

Recommendations

Prenatal Assessment and Planning

Almost two-thirds of CDH cases are identified prenatally through routine ultrasound monitoring.[2,3] Following a diagnosis of CDH, a comprehensive assessment is critical to provide prognostic information, stratify risk, and inform interdisciplinary prenatal planning. The following assessments are important components of risk stratification[2,4-8]:

- Amniocentesis:
  - Microarray analysis to investigate the presence of aneuploidy. Single gene disorders cannot be ruled out.
- Targeted ultrasound:
  - Ascertain the presence of other congenital abnormalities (including cardiac as the most common one affecting prognosis)
  - Observed to expected lung (area) to head (circumference) ratio (O/E LHR)
    - Helps predict survival and the severity of pulmonary hypoplasia
    - An O/E LHR < 25% in left-sided CDH predicts poor outcome; in right-sided CDH, an O/E LHR < 45% may predict poor outcome
  - Position of the stomach in the thorax
  - Presence of liver herniation
  - Presence of a hernia sac
- Fetal MRI:
  - Observed to expected total fetal lung volume (O/E TFLV)
    - O/E TFLV <25% is associated with greater likelihood of mortality
  - Percent Predicted Lung Volume (PPLV)
    - PPLV <15% is associated with reduced survival
  - Presence and degree of liver herniation
  - Presence of other congenital abnormalities
- Fetal Echocardiogram:
  - 20-22 weeks:
    - Evaluate for associated congenital cardiac defect and for physiologic and structural consequence of cardiac shift and compression
  - 32-36 weeks (optional):
    - Evaluate for potential prognostic markers (i.e. hyperoxia testing, branch PA size, and flow and interval growth of cardiac structures)

Following a comprehensive assessment of the above factors, multidisciplinary prenatal consultation is completed to review and determine the most appropriate options to optimize outcomes for mother and baby in a manner that is consistent with the family's wishes. Components of this consultation include planning for the following[2,3,6-8].

- Consideration for fetoscopic endotracheal occlusion (FETO)
  - The idea behind FETO is that tracheal occlusion may induce pulmonary growth. Early studies suggest a potential survival benefit associated with this intervention in severe CDH with O/E TFLV < 25%. Preterm delivery is a potential adverse effect which must be considered.
- Potential need for ECMO
  - Babies with O/E TFLV <25%, PPLV <15%, O/E LHR < 25%, and/or liver up are more likely to require ECMO support
- Mode of delivery

- Timing of delivery
- Delivery location
    - Meriter
    - AFCH
    - UH (high maternal risk)
- Newborn genetic testing
    - Hold cord blood for microarray (sodium heparin tube) and/or genomic testing (lavender tube) based on prenatal or postnatal guidance from genetics

Delivery Room Management and Initial stabilization

Delivery room and early newborn care should focus on establishing stability on the ventilator, providing medication for sedation and pain control, decompressing the bowel, and placement of venous and arterial access.

Key management points include:

1. Avoid bag-mask ventilation, as it may cause distension of the stomach which limits lung expansion.[4,9,10] *(UW Health Very Low quality evidence, Strong recommendation)*
    a. If bag-mask is required, cricoid pressure should be used.

2. Orotracheal intubation should occur immediately after birth with rare exception (i.e. low risk and without respiratory distress), and should be performed by an experienced provider (e.g., senior fellow, attending using a cuffed ET tube if available).[4,9,10] *(UW Health Very Low quality evidence, Strong recommendation)*
    a. Initiate NeoPuff (PEEP 5 and PIP no more than 25 cm H20).

3. Perform gastric decompression immediately to allow for lung expansion[9]. *(UW Health Very Low quality evidence, Strong recommendation)*
    a. Use10F Replogle (or the largest that will fit) and 30 ml syringe in the delivery room.

4. As soon as possible, give IV morphine 0.05-0.1 mg/kg[9]. *(UW Health Very Low quality evidence, Strong recommendation)*

5. Do not perform delayed cord clamping due to need for immediate intubation. (*UW Health Very Low quality evidence IEO, Conditional recommendation*).

Initial Stabilization

Lines/Drains/Tubes/Initial Labs

1. The Replogle should be placed to low intermittent suction immediately.

2. Umbilical catheter access should be placed on arrival to the NICU.
    a. Umbilical venous catheter (UVC) should be double-lumen
    b. Send arterial blood gas with Hgb, lactate, glucose, and blood culture
    c. Placement of the UAC and UVC should be confirmed by x-ray, however a UVC may also be confirmed using an ultrasound or echocardiogram, as x-ray can be challenging to interpret due to malposition of the liver.[10]

Analgesia and Sedation

1. Analgesia and sedation should be provided and monitored with validated scales[4,9,10] (i.e., State Behavioral Scale (SBS) or Neonatal Pain Agitation and Sedation Scale (N-PASS)). *(UW Health Very Low quality evidence, Strong recommendation)*
    a. Initiate dexmedetomidine first line.
    b. Morphine should be utilized, as needed for additional analgesia, with the goal of using the minimal necessary dose.
    c. Midazolam can be considered for additional sedation, if clinically necessary (after optimizing dexmedetomidine and morphine dosages).

2. Paralysis and deep sedation should generally be avoided; may consider for patients requiring maximal respiratory support.[2,4,9,10] *(UW Health Low quality evidence, Conditional recommendation)*
    a. Deep sedation using midazolam and morphine is required when administering paralysis.

Antibiotics

1. Routine antibiotics are not recommended for all infants with CDH; initiation should be based on clinical indication. *(UW Health Very Low quality evidence, Conditional recommendation)*
    a. Due to the increased risk for ototoxicity and nephrotoxicity with gentamicin, ampicillin and cefotaxime (or ceftazidime) are recommended. (*UW Health Very Low quality evidence IEO, Conditional recommendation*).

Initial Ventilator Management

1. Position the patient with the affected side slightly down and the head turned toward the defect to optimize inflation of “good lung”. *(UW Health Very Low quality evidence IEO, Strong recommendation)*

2. Pre-ductal saturations should be monitored routinely. Post-ductal saturation monitoring is optional. *(UW Health Good practice statement)*
    a. In the first 1hr, target pre-ductal saturations ≥ 75% [9].

3. A “gentle ventilation” strategy using conventional mechanical ventilation is recommended .[4,9,10] *(UW Health Low quality evidence, Conditional recommendation)* This strategy involves tolerance of hypercapnia in order to minimize exposure to high airway pressures and reduce the risk of ventilator induced lung injury. Pressure control modes of ventilation are generally preferred. The following initial ventilator settings can be used[9,10]:
    a. PIP 15 (Max 25) cm H_2O
    b. PEEP 3 (Max 6) cm H_2O
    c. Rate 40 breaths/minute (Max 60)
    d. iT 0.3 - 0.4
    e. FiO2 1.00. Wean FiO2 slowly for pre-ductal saturation > 95%

4. Routine use of surfactant is not recommended, but can be considered in premature infants.[9-12] *(UW Health Low quality evidence, Conditional recommendation)*

Imaging

1. Consider delaying initial ECHO for ~24hrs during the period of initial stabilization provided infant is clinically stable, to minimize stimulation. Obtain within the first 48 hours if feasible noting the following considerations[4]. *(UW Health Very Low quality evidence, Conditional recommendation)*

a. If fetal ECHO was performed and anatomy was well defined, there is reduced urgency to obtain ECHO early on.
b. Limited ECHO for PPHN may be considered.

2. Head ultrasound should be considered if ECMO is likely[13]. *(UW Health Very Low quality evidence, Conditional recommendation)*
    a. Consider delaying cranial ultrasound until after transfer to AFCH if clinically stable.

Admission Location and Transfer Considerations

1. Babies born at AFCH or UH will be admitted to the NICU for further stabilization and management. *(UW Health Very Low quality evidence – IEO, Strong recommendation)*
2. For babies born at Meriter, transfer to AFCH NICU should occur within the first 12 hours of life. *(UW Health Very Low quality evidence – IEO, Strong recommendation)*
3. Independent of birth location, if there is imminent concern the infant will require ECMO, further discussion with NICU, PICU, and pediatric surgery will guide subsequent decisions.

Preoperative Management

Nursing Care and Room Setup

1. When possible, position the ventilator on the patient's left side. It is preferred to position head of the patient at the foot of the bed for airway accessibility.

2. Birth weight is used as the dosing weight for at least 7 days. Weight should be measured on admission and then should not be routinely performed but can be obtained when volume status is uncertain.

3. Minimal stimulation practices should be followed, including the following: *(UW Health Good practice statement)*
    a. Complete physiologic assessments should be performed every 4-6 hours, with focused assessments as needed. [9]
        i. Routine pupil checks should be completed only on admission and with acute changes.
        ii. Temperature measurements should NEVER be obtained rectally; perform axillary measurements with full assessments.
        iii. When continuous arterial blood pressure monitoring is available check cuff pressures only once per shift to assess accuracy.
    b. Apply eye covers and earmuffs.
    c. Coordinate hands on assessments and cluster care to minimize disruptions to the infant.
    d. Minimal repositions should be performed to relieve pressure points only. Holding is not allowed until the patient is stable post-operatively.

4. Standard closed suctioning should be performed as clinically indicated. If unable to resolve clinical decompensation following closed suctioning, consider open bag suctioning. (*UW Health Good practice statement*)

5. Monitoring of the following parameters is recommended (*UW Health Good practice statement*):
    a. Cerebral and renal NIRS
    b. Pre- and post-ductal saturations
    c. aEEG monitoring for paralyzed infants
    d. Continuous arterial blood pressure monitoring

Lines/Drains/Tubes

1. Obtain daily babygram to ensure proper positioning of all lines and tubes, as well as adequate bowel decompression. *(UW Health Good practice statement)*
2. Replogle must be placed to low-intermittent suction and in good position.
    a. Use 10 French catheter in patients > 36 weeks gestational age.
        i. If decompression inadequate due to malpositioning, consider downsizing.
3. If the surgical team plans to remove umbilical access in the OR, a double-lumen PICC should be placed pre-operatively.

Ventilator Management

Gentle ventilation and permissive hypercapnia (involving the use of lower tidal volumes and higher respiratory rates) should be continued as a cornerstone of therapy. Minimizing the risk of oxygen toxicity is also an important goal. Below, we provide target ranges for ventilation and oxygenation, temporary allowable excursions from target ranges, and specific strategies to consider as a part of a gentle ventilation strategy. Indications for increased support and suggested escalation strategies are also outlined. [2-4,9,10,14-16]

Monitoring and Target Ranges[4,9,10,14,15]

1. Ventilation
    a. Permissive hypercarbia should be tolerated as long as the patient is comfortable and pulmonary hypertension is adequately controlled.
    b. Target ranges *(UW Health Low quality evidence, Strong recommendation)*
        i. pH ≥ 7.25 and PCO_2 45-65 mmHg
        ii. Higher PCO_2 levels may be tolerated during the initial transition period and intermittently during the pre-operative period.
        iii. Avoid rapid changes in PCO_2, as this may result in acute changes in cerebral blood flow.
2. Oxygenation
    a. Optimizing cardiac output and oxygen carrying capacity may allow patients to better tolerate lower saturations during initial transition and pre-operative period.
    b. Target pre-ductal saturations as follows: *(UW Health Low quality evidence, Strong recommendation)*
        i. In the first 1 hr of life: ≥75%
        ii. 1-2 hrs of life: 80-90%
        iii. By 3 hrs of life: >90%
        iv. Wean FiO_2 slowly for pre-ductal saturation > 95% (by 2-5% every 30-60 minutes) to minimize reactive pulmonary hypertension.
            1. In the first 24 hrs of life, do not wean FiO2 below 30%.
            2. Beyond 24 hrs of life, if stable on FiO2 0.3 (i.e. PaO2 remaining >100 and pre-ductal saturation 99-100%), consider weaning FiO2 further.
    c. Post-ductal saturations (if being monitored) as low as 70 may be acceptable (and even lower if urine output remains reasonable and lactate low). *(UW Health Low quality evidence, Conditional recommendation)*
        i. Post-ductal saturations are indicative of right-to-left shunt and not a measure of cerebral or myocardial oxygen delivery.
        ii. Post-ductal monitoring is primarily helpful to understand and monitor lability of PVR rather than an indicator of O_2 content.

d. PaO_2 measurements
   i. Should *not* be the sole indicator for increasing support. More reliable
      a. measures include trending saturations and evidence of end organ function.
   ii. Right radial arterial monitoring may be useful if there is a large right-to-left ductal shunt (to help avoid cerebral and retinal hyperoxygenation) but is not necessary for acute monitoring.
e. Cerebral and renal NIRS, lactate, urine output, and venous saturations are helpful additional indicators of systemic oxygen delivery and should be monitored. *(UW Health Good practice statement)*

Indications for Increasing Support and Approach [4,9,10,15]

1. Most CDH patients have increased work of breathing at baseline with tachypnea and mild retractions being common. Prior to increasing respiratory support it is important to assess the following (*Good practice statement*):
   a. changes in respiratory pattern from baseline
   b. adequacy of sedation
   c. position of patient
   d. position of ETT
   e. need for suctioning
   f. adequacy of bowel decompression.
2. When sustained, the following are reasons to consider ventilator changes:
   a. Paradoxical chest wall movement
   b. Significant increase in retractions
   c. Preductal SaO_2 < 80%
   d. pCO_2 > 65 mmHg (pH < 7.25)
   e. Sustained pre-ductal to post-ductal SaO_2 difference >10% indicate PHTN and should prompt investigation (see pulmonary hypertension section).
3. Ventilator management considerations for hypoxemia or hypercarbia
   a. SIMV-PC considerations
      i. Focus on changes that minimize exposure to high pressure (i.e., preference for high rate over high pressure [PIP > 25]).
      ii. IMV rate as high as 60 for respiratory distress
         1. Decrease PEEP to avoid air trapping
         2. Decrease iT 0.3 – 0.4 while monitoring I:E ratio
      iii. Consider decreasing PEEP to increase ΔP while minimizing exposure to high pressure. Before increasing PIP, evaluate compliance and pressure-volume loops.
   b. Consider high frequency oscillatory ventilation (HFOV) in the following situations[9,14,15] *(UW Health Very Low quality evidence, Conditional recommendation)*:
      - Hypercapnia not improved on conventional ventilation (PIP > 25 cm H_2O and Rate > 60) while maximizing sedation, positioning, and pulmonary toilette
      - Failure to oxygenate despite use of conventional ventilation in conjunction with iNO
      - For pneumothorax
         i. The following initial HFOV settings are recommended:
            1. Mean airway pressures (MAP) 14 – 16 cm H_2O
            2. Hz 10 – 12. This range is higher than normal for a term infant but CDH patients have pulmonary hypoplasia and therefore higher Hz is preferred for lung protection and is generally adequate.

3. Amplitude 30 – 40 cm H_2O
4. Goal rib expansion: 8 – 10 ribs on contralateral side. To minimize lung injury, avoid flattening of the diaphragm.

i. Obtain a babygram within 30 min of transition to HFOV to evaluate for hyperinflation.

iii. Closely monitor for signs of hypotension due to decreased venous return.

Weaning

1. Weaning in the pre-operative period should be very gradual to avoid the need to dramatically increase support during periods of decompensation.
2. Prioritize weaning pressure over FiO_2 and rate. *(UW Health Very Low quality evidence, Strong recommendation)*

Acute Pulmonary Hypertension (PHTN) and Hemodynamic Support

Some PHTN is expected; right (systolic) and left (diastolic) heart function are the primary concerns. In the setting of PHTN with good right ventricular (RV) function and pre-ductal saturation, there is likely no need to treat. Furthermore, the systemic circulation may be dependent on ductal dependent perfusion from the RV given *in utero* underdevelopment and diastolic dysfunction of the LV.

Initiation of Treatment for Pulmonary Hypertension

1. Treatment for acute PHTN is recommended in patients with one or more of the following indications[4,9,14,17,18] *(UW Health Low quality evidence, Strong recommendation)*:
    a. Pre-ductal SpO_2 < 90% despite optimizing other ventilatory maneuvers
    b. Oxygenation index (OI) > 25
    c. Post-ductal saturations < 70% and/or evidence of end organ dysfunction (e.g., rising lactate, acidosis, or oliguria)
    d. Echocardiogram demonstrates systemic or near-systemic right-sided pressures with depressed RV function or shows evidence of supra-systemic to near-systemic right-sided pressures with borderline oxygenation.

Pulmonary Hypertension Treatment Options

1. A trial of inhaled nitric oxide (iNO) should be initiated as the first line treatment for suspected pulmonary hypertension; if there is evidence of significant LV dysfunction, have caution as iNO may worsen hemodynamic status[4,9,17-22]. *(UW Health Very Low quality evidence, Conditional recommendation)*
    a. Use starting dose of 20 ppm.
    b. Reassessment should occur within 30-60 min of iNO initiation. If no improvement, may consider increasing dose to 40. If still no response to iNO, consider weaning off iNO quickly (i.e. over 60 minutes).
    c. Monitor methemoglobin level (normal < 1.2%) daily while receiving iNO. *(UW Health Good Practice Statement)*
2. For RV dysfunction associated with pulmonary hypertension, milrinone (0.5 mcg/kg/min) is the preferred first line agent.[2,4,23,24] *(UW Health Very Low quality evidence, Conditional recommendation)*
    a. In patients with hypotension and RV dysfunction receiving milrinone, vasopressor support is needed.

3. Prostaglandins may be useful in the specific circumstances of severe PHTN.[4,9,19,22,25] *(UW Health Very Low quality evidence, Conditional recommendation)*
    a. In babies with pulmonary hypertension and RV failure or in the case of a closing ductus, intravenous PGE1 can be used to open the ductus arteriosus and reduce RV afterload.
    b. Inhaled prostacyclin has the advantage of possibly having improved ventilation-perfusion matching and may be useful in the acute treatment of PHTN. For dosing recommendations related to inhaled prostacyclin (epoprostenol), see UW Health Epoprostenol Inhaled – Neonatal/Pediatric/Adult – Inpatient Guideline.
4. Sildenafil may be considered in cases of refractory pulmonary hypertension (typically late or subacute phase) or if longer-term treatment of pulmonary hypertension is necessary[4,9,15]. (*UW Health Low quality evidence, Conditional recommendation*)
    a. Start sildenafil dose at 0.5mg/kg Q6hrs (max dose of 1mg/kg Q6hrs). Clamp NG/OG tube x 30min following dose.

Hemodynamic Support

1. If there is evidence of poor end organ perfusion and/or hypotension, treatment considerations include the following[4,9,10] *(UW Health Very Low quality evidence, Conditional recommendation)*:
    a. For hypovolemia, use isotonic crystalloids judiciously (10-20 mL/kg).
    b. For hypotension, use of the following medications is encouraged to increase SVR and raise systemic blood pressure in the setting of high PVR and good RV function in an effort to decrease right-to-left ductal shunting.[9,10]
        - Norepinephrine (suggested dosing range 0.05-1.0 mcg/kg/min)
        - Epinephrine (suggested dosing range 0.01-0.1 mcg/kg/min)[26-28]
        - Vasopressin (suggested starting dose 0.17-10 milliUnits/kg/min)
        - Dopamine may worsen pulmonary hypertension due to pulmonary vasoconstrictive effects and should be considered with caution in this context (suggested dosing range 5-25 mcg/kg/min).
    c. For ventricular dysfunction, dobutamine (suggested dosing range 2-20 mcg/kg/min) is an additional therapeutic option.
    d. Hydrocortisone (1 mg/kg Q8 hrs) may be considered as an adjunct agent to address adrenal insufficiency and hypotension.

2. Consider cardiology consult when >2 systemic vasodilators (not including iNO) are required. (*UW Health Good practice statement*)

Extracorporeal membrane oxygenation (ECMO)

1. There are no specific criteria for ECMO in CDH patients; however the following indications and contraindications may be used collectively to determine eligibility[8,9,15,29,30] *(UW Health Low quality evidence, Conditional recommendation)*. If baby is meeting criteria for ECMO consideration, early discussion with NICU, PICU and pediatric surgery attendings should occur.

Table 1. Postnatal Considerations and Contraindications for ECMO [1,8,9,15,29-32]

Criteria for ECMO Consideration	Absolute ECMO Contraindications	Relative ECMO Contraindications
When sustained and refractory to treatment: o Preductal SaO_2 < 85% o PaCO2 >70 and/or pH <7.20 o OI > 40† o PIP > 30 o HFOV AMP > 45 o Lactate (arterial) > 4 mmol/L o Circulatory failure ▪ Severe hypotension ▪ LV failure ▪ Severe PHTN resulting in RV failure	• Gestational age < 32 weeks • Birth weight < 1.6 kg • Lethal comorbidity (Pentalogy of Cantrell, Frynn's syndrome, Bilateral defects) • IVH ≥ grade 3	• Gestational age 32-34 weeks • Birth weight < 2 kg • IVH grade 2 • Cardiac lesion (coarctation)** • Failure to achieve SaO_2 > 85% (and sustain for 1 hour) within the first 2-4 hours of life despite maximal ventilatory maneuvers

** ECMO for patients with CDH and congenital heart disease will be considered on a case-by-case basis depending on the type of cardiac lesion and severity of the CDH.

† $OI = (FiO_2 \times M_{PAW}) / PaO_2$

2. Veno-venous (VV) ECMO may be considered if cardiac function is adequate, even in the setting of pulmonary hypertension, as the oxygenation of venous blood can mitigate the effects of right-to-left shunt.[15]

Other management considerations for babies requiring ECMO are provided in the ECMO: Initiation and Management - Pediatric/Neonatal – Inpatient/Emergency Department - Clinical Practice Guideline.

Surgery

General Philosophy and Timing

1. The timing of surgery and approach to surgical repair should be based upon each individual clinical scenario, risks and benefits of the procedure, and clinical judgement of the medical and surgical teams. *(UW Health Low quality evidence, Strong recommendation)* A summary of surgical considerations includes the following[4,8,9]:
   a. Timing of repair
      i. Infants not requiring ECMO should undergo repair after clinical stabilization *(UW Health Very Low quality evidence, Conditional recommendation)*. Physiologic criteria to consider when evaluating stability include:
         • Oxygenation Index (OI) < 9
         • urine output > 1 mL/kg/h
         • FiO2 < 0.5
         • preductal oxygen saturation between 85% and 95%
         • normal mean arterial pressure for gestational age
         • lactate < 3 mmol/L
         • estimated pulmonary artery pressures less than systemic pressure.
      ii. However, failure to meet these criteria does not preclude surgical repair.
      iii. For infants requiring ECMO, the evidence regarding timing of surgical repair is controversial.
         ▪ There may be benefit to delaying repair until after decannulation for infants in whom decannulation is possible.

- If the infant has a very severe CDH phenotype, it is possible that early (<72hrs) repair on ECMO may have benefits.

b. Approach to surgical repair
    i. Minimally invasive (thoracopic) techniques have been used although potential benefits of this approach as compared to open repair have not been established. Thoracopic repairs have been found to be associated with greater recurrence rates and can lead to hypercarbia and acidosis, which may have important physiologic consequences.
        - For thoracoscopic repairs, close monitoring of intraoperative PaCO2 and pH is warranted.
    ii. For small defects, primary surgical closure may be performed. For large defects not amenable to primary closure, options include closure with oversized prosthetic material or abdominal wall muscle flap.

Postoperative Care

Respiratory Considerations

1. Patients with CDH will often have lower tidal volumes and higher respiratory rates. The goal is to achieve near-normal ventilation and oxygenation.
2. Pulmonary toilette should begin with suctioning post-operatively. Open bag-suctioning may be required, particularly in the immediate post-operative period.
3. Pleural effusion on the affected side is an expected consequence of repair of CDH. However, if there is evidence of mass effect (i.e., shift of the mediastinum to contralateral side, or decreased respiratory compliance) consider drainage and investigate etiology (e.g., chylothorax).

Post-operative Ventilator Management

1. Because of the increased need for sedation and risk of PHTN, patients generally are not weaned for 48 – 72 hours post-operatively unless over-ventilated (CO2 < 45), over oxygenated (saturation > 98%), or receiving large tidal volumes (more than 6 mL/kg or more than pre-operatively).
2. Neurally adjusted ventilator assist (NAVA) is the preferred mode of ventilation to improve patient synchrony while minimizing sedation[33,34]. *(UW Health Very Low quality evidence, Conditional recommendation)*
    a. Establishing NAVA
        i. X-ray confirmation of appropriate NAVA catheter placement is necessary prior to initiation.
        ii. Prior to initiation of NAVA, it is important to evaluate peak pressures, tidal volumes and Edi signal, as this well help determine the starting NAVA level.
    b. Recommended initial settings
        i. NAVA level of 1-1.5 (adjust by increments of 0.1-0.2 to obtain appropriate peak pressures and tidal volumes)
        ii. PEEP of 5 cm H_2O (range 3-7)
        iii. Back up setting should be set (i.e., PEEP 5 cm H_2O, PIP 20 cm H_2O, Rate 40)
    c. Indications for weaning:
        i. NAVA level is maintained and tidal volumes improve with decreasing/stable Edi values and peak pressures

ii. NAVA level is weaned by 0.1-0.2 and patient is able to maintain appropriate tidal volumes without significant increase in work of breathing.

3. When NAVA mode of ventilation is not feasible (deep level of sedation required, inadequate Edi signal, or other clinical considerations), conventional modes of ventilation may be used.
    a. Conventional ventilator mode considerations:
        i. Higher rates are preferred to higher peak pressures to maintain pH > 7.3.
        ii. If peak pressures > 25 to generate goal tidal volumes (from pre-op), consider trial of decreased PEEP (as low as 4 cm H2O) OR transition to HFOV.
    b. Weaning from conventional modes of ventilation
        i. Increased pressure support may be useful to allow weaning of rate and enhanced patient synchrony.
        ii. Inspiratory pressures should be weaned (if possible) if chest excursion good and reasonable tidal volumes.
        iii. Once peak pressures in low 20s weaning rate is the next priority.
4. When patient is ready to extubate, it is recommended to extubate to either NIV-NAVA or NIPPV *(UW Health Very Low quality evidence IEO, Conditional recommendation)*.
    a. NIV-NAVA
        i. Consider setting the NIV-NAVA level 1 above the NAVA level used prior to extubation.
    b. NIPPV
        i. Consider the following settings: PIP 20 – 24 cm H2O, PEEP 5 – 6 cm H2O, Rate 40, I-time 0.5.

Pulmonary Hypertension

1. Patients with pulmonary hypertension prior to surgery should be monitored for pulmonary hypertension and its sequalae post-operatively with an echocardiogram *(UW Health Good practice statement)*.
2. Weaning of inhaled nitric oxide should occur coincidently with ventilator weaning. It is recommended to begin weaning iNO once FiO_2 is < 0.4. *(UW Health Very Low quality evidence IEO, Conditional recommendation)*
    a. A repeat echocardiogram should be obtained once the patient is off iNO
    b. Consider weekly echocardiogram while weaning respiratory support.
3. Patients who cannot be effectively weaned from iNO should be evaluated by interventional cardiology to consider direct measurement of the pulmonary vascular pressures and responsiveness to pulmonary vasodilating medications. Long-term pulmonary vasodilator treatment might include sildenafil, prostacyclin (IV or inhaled) or endothelin receptor antagonists.[4,9,15,16] Caution must be used as all these (except inhaled prostacyclin) can worsen V/Q mismatch in CDH patients who are hypoxic. *(UW Health Low quality evidence, Conditional recommendation)*
    a. Consider cardiology consult for babies requiring vasodilator therapy beyond 4 weeks post-op. (*UW Health Good practice statement*)

Gastrointestinal

1. Babies with CDH are at significantly increased risk of reflux and the sequalae (e.g. aspiration, malnutrition) can be significant. If symptoms exist, consider the following[3,9]:
    - Extending feeding times liberally (inclusive of continuous feeding, if needed) to minimize the need for ongoing TPN/IL
    - Initiate reflux treatment (H2 blocker or PPI)
    - Post-pyloric feeding

- Ruling out malrotation when symptoms persist
  (*UW Health Very Low quality evidence, Conditional recommendation*)

Discharge Planning

1. The following imaging studies are recommended prior to discharge if not already completed within 2 weeks of the discharge date*(UW Health Very Low quality evidence IEO, Strong recommendation)*:[35,36]
    a. Echocardiogram and chest x-ray (for babies requiring home oxygen)
    b. Brain MRI (ideally at 1-2 months of age) should be obtained in any newborn with:
        - multiple congenital anomalies
        - prematurity
        - need for fetal surgery
        - need for ECMO

Disclaimer

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Conflicts of Interest

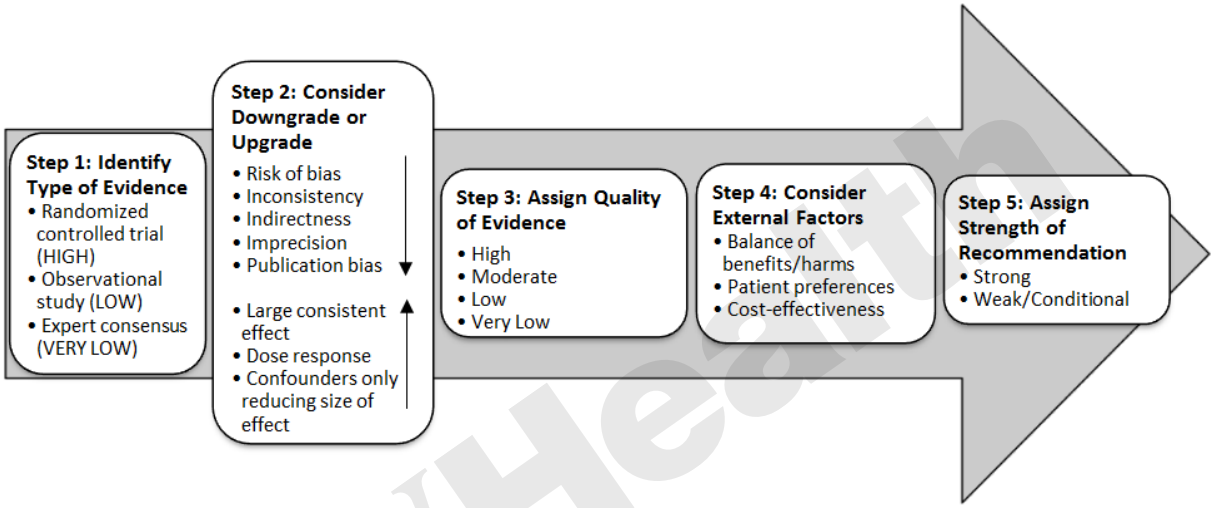
All guideline workgroup members are expected to follow institutional policies and procedures around conflicts of interest. Actions in which a guideline member discloses a conflict of interest relevant to the guideline topic may include, but is not limited to, abstaining from voting, dismissal during comment and voting period, or recusal from requesting and/or participation in the decision-making process.

Methodology

Development Process

Each guideline is reviewed and updated approximately every 3-5 years, but frequency will vary in consideration of the primary literature and relevant practice changes. All guidelines are developed using the guiding principles, standard processes, and styling outlined in the UW Health Clinical Practice Guideline Resource Guide. This includes expectations for workgroup composition and recruitment strategies, disclosure and management of conflict of interest for participating workgroup members, literature review techniques, evidence grading resources, required approval bodies, and suggestions for communication and implementation.

GRADE Methodology adapted by UW Health



Rating Scheme for the Strength of the Evidence/Recommendations:

GRADE Ranking of Evidence

High	We are confident that the effect in the study reflects the actual effect.
Moderate	We are quite confident that the effect in the study is close to the true effect, but it is also possible it is substantially different.
Low	The true effect may differ significantly from the estimate.
Very Low	The true effect is likely to be substantially different from the estimated effect.
Very Low (Internal Expert Opinion)	The true effect is likely to be substantially different from the estimated effect. This category of recommendation for or against a specific intervention is derived strictly from the expert opinions of UW Health healthcare professionals with experience in the relevant specialty(ies). This is used in the absence of published evidence or external opinion addressing the specific intervention.

GRADE Ratings for Recommendations For or Against Practice

Strong (S)	Generally, should be performed (i.e., the net benefit of the treatment is clear, patient values and circumstances are unlikely to affect the decision.)

Conditional (C)	Recommendation may be conditional upon patient values and preferences, the resources available, or the setting in which the intervention will be implemented.
Good Practice Statement	Generally, should be performed (i.e., the expected benefit of the treatment is substantial, expected costs or risk are minimal, and patient values and circumstances are unlikely to affect the decision.) This classification is used for recommendations that guideline members feel are important and for which there is uniform support, but for which evidence directly assessing the specific intervention (or practice) is not available and is highly unlikely to ever be studied (because it may not be warranted or feasible). Such recommendations may have strong indirect evidence of support

Recognition of Potential Health Care Disparities: Hospital survival for congenital diaphragmatic hernia was related to sex, birth weight, race and socioeconomic status in a retrospective analysis of 2,774 hospitalizations using the Kids' Inpatient Database.[37] Higher survival rates were seen in males, patients with a birth weight $\geq$ 3 kg, white patients, patients with private insurance, and patients in the highest median household income quartile. Black race and other non-Hispanic minorities were identified as independent predictors of mortality. Clinical staff and providers should remain aware of these statistics, and work to provide the best appropriate care to all patients, regardless of gender, race, or socioeconomic status.

Collateral Tools & Resources

Guideline Metrics:

- Mortality rate
- Average timing of repair
- Percentage of CDH patients requiring ECMO
- Percentage of CDH patients with imaging studies completed prior to discharge

Policies
Nursing Patient Care Policy 1.48-P- Care of Umbilical Catheters (Arterial and Venous)
Nursing Patient Care Policy 1.49-P- Use of Near-Infrared Spectroscopy (Cerebral and Somatic Oximetry) in the Pediatric and Neonatal ICU
Nursing Patient Care Policy 2.20- Care and Maintenance of Enteral Tubes
Nursing Patient Care Policy 7.11-P- Care of the Intubated Patient
Respiratory Care Services Policy 1.53 (AFCH Initial Ventilator Management and Weaning Algorithms)
Respiratory Care Services Policy 2.02- Mechanical Ventilation Adult and Pediatric
Respiratory Care Services Policy 2.03- High Frequency Oscillatory Ventilation (HFOV)
Respiratory Care Services policy 3.42- Suctioning
Respiratory Care Services policy 3.53- Nitric Oxide
UW Health Clinical Policy 2.3.31- Administration of Non-Invasive Ventilation (Commonly referred to as BiPaP and NIV)

Procedures (Elsevier Skills)
Chest Tube: Closed Drainage System (Neonatal)

Patient Resources
Kids Health- Genetic Counseling
Kids Health- Birth Defects

Practice Protocols
Pain and Agitation Continuous Infusion Titration – Pediatric – Inpatient [5]

Collateral UW Health Guidelines
Enteral Nutrition – Neonatal – Inpatient Guideline
Epoprostenol Inhaled – Adult/Pediatric/Neonatal – Inpatient Guideline
Extracorporeal Membrane Oxygenation (ECMO): Initiation and Management - Pediatric/Neonatal – Inpatient/Emergency Department Guideline
Neonatal Analgesia – Neonatal – Inpatient/Ambulatory Guideline
Parenteral Nutrition – Pediatric/Neonatal – Inpatient/Ambulatory Guideline
Prevention of Ventilator Associated Pneumonia – Pediatric/Neonatal – Inpatient Guideline

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