

Children's Mercy Kansas City

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Clinical Pathways

Evidence-Based Practice Collaborative

9-2024

Congenital Diaphragmatic Hernia

Children's Mercy Kansas City

These guidelines do not establish a standard of care to be followed in every case. It is recognized that each case is different and those individuals involved in providing health care are expected to use their judgment in determining what is in the best interests of the patient based on the circumstances existing at the time. It is impossible to anticipate all possible situations that may exist and to prepare guidelines for each. Accordingly, these guidelines should guide care with the understanding that departures from them may be required at times.

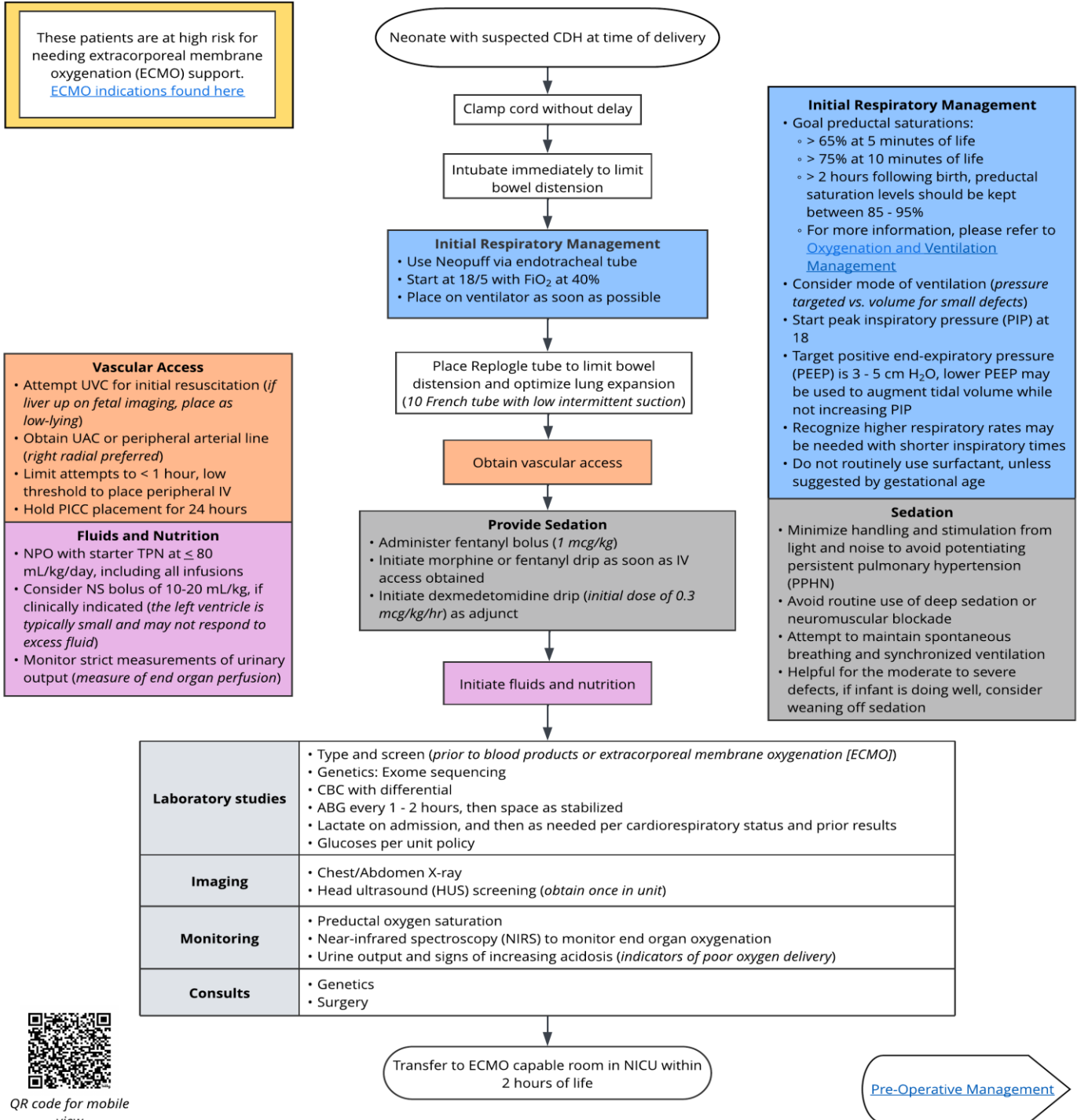
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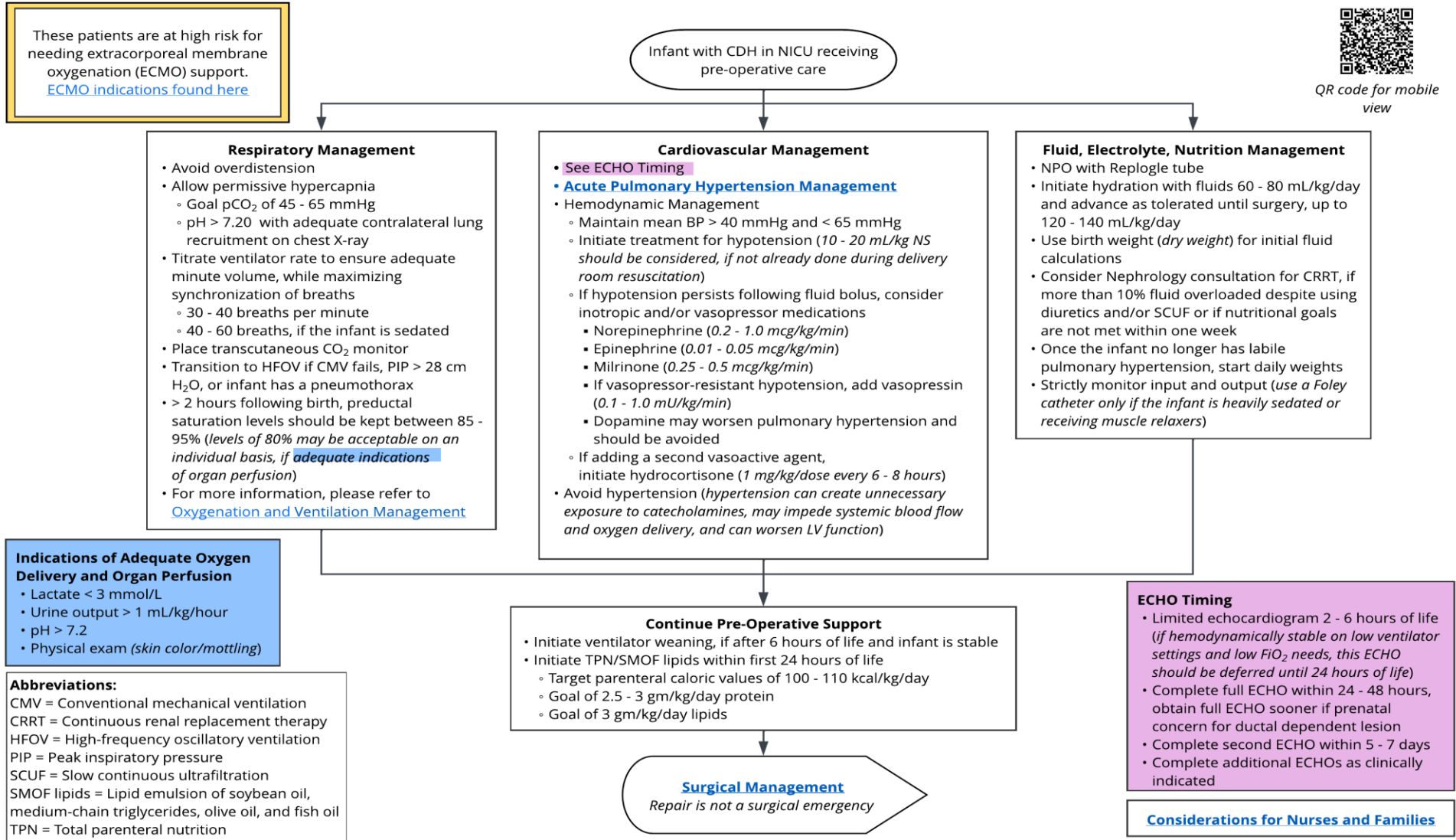
Congenital Diaphragmatic Hernia Clinical Pathway Synopsis

Congenital Diaphragmatic Hernia (CDH): Delivery Room Resuscitation Algorithm



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Congenital Diaphragmatic Hernia (CDH): Pre-Operative Management Algorithm



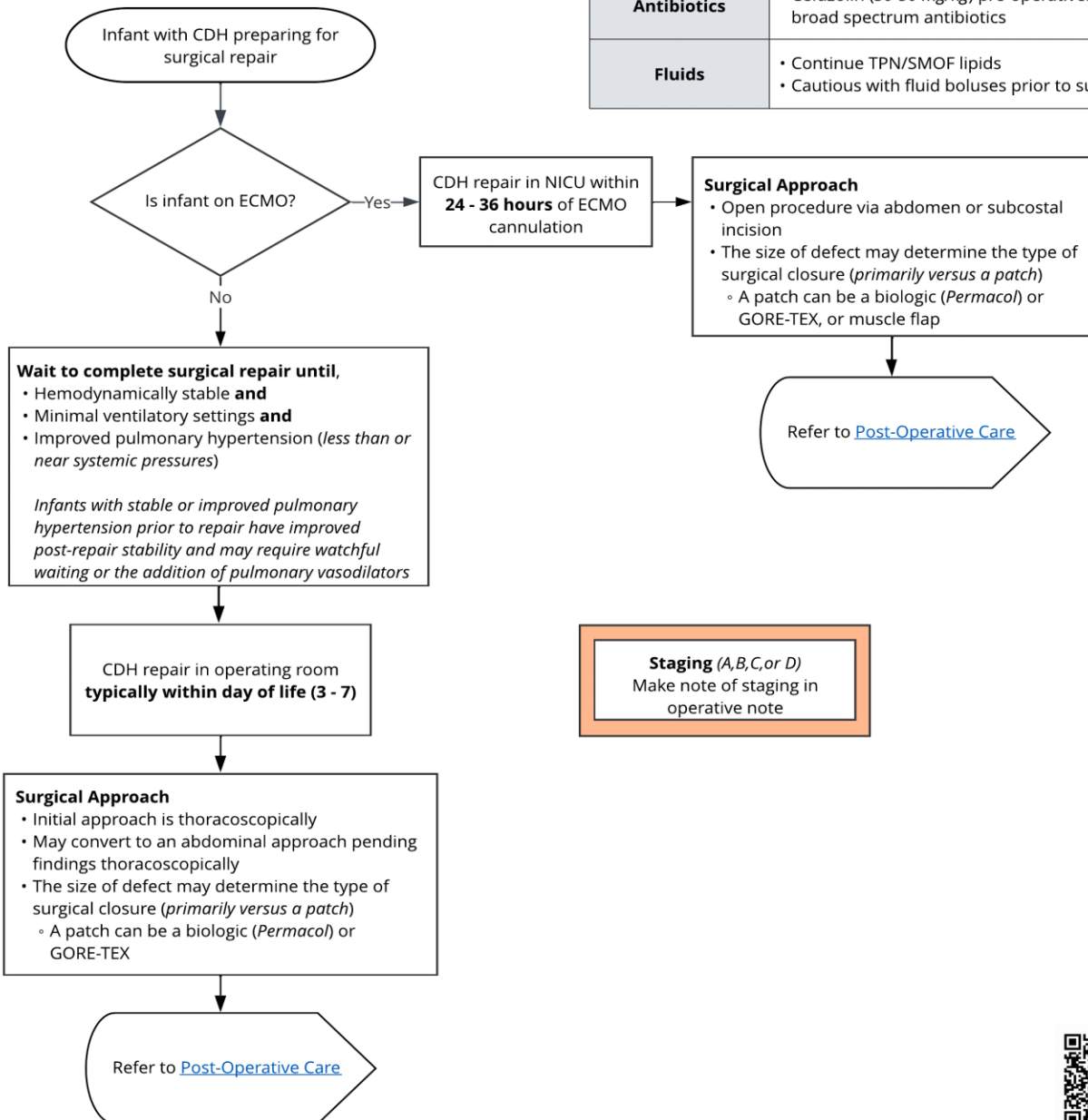
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Congenital Diaphragmatic Hernia (CDH): Surgical Management Algorithm

Line Management Prior to Surgery

- Conversion of UVC to PICC
- Attempt to convert UAC to a peripheral arterial line

Preparing for Surgery	
Laboratory studies	<ul style="list-style-type: none"> • CBC within 24 hours of surgery • Coagulation studies within 24 hours of surgery • ABG
Vascular access	<ul style="list-style-type: none"> • PICC line • Peripheral arterial line
Antibiotics	<ul style="list-style-type: none"> • Cefazolin (30-50 mg/kg) pre-operatively unless on broad spectrum antibiotics
Fluids	<ul style="list-style-type: none"> • Continue TPN/SMOF lipids • Cautious with fluid boluses prior to surgery



Staging (A,B,C,or D)
Make note of staging in operative note



QR code for mobile view

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Congenital Diaphragmatic Hernia: Post-Operative Care Algorithm

Abbreviations:
 OT = Occupational therapy
 PEEP = Positive end-expiratory pressure
 PO = By mouth



QR code for mobile view

Infant following surgical repair of CDH

Special Considerations

- [Indications for extracorporeal membrane oxygenation \(ECMO\)](#) do not change in the post-operative period
- Continue [Pulmonary Hypertension Management](#)
- If chest tube present, leave to water seal, **not** suction
- Continue near-infrared spectroscopy (NIRS) monitoring until acidosis is resolved and infant has normal urine output
- Pleural effusion is an expected sequelae of repair

Post-Operative Ventilatory Management

- Pulmonary toileting with mucolytic should be considered after 48 hours following repair, pending clinical stability.
- May need to increase ventilatory support in the immediate post-operative period to regain stability; however the objective remains gentle ventilation
- Continue permissive hypercapnia CO₂ 45 - 65 mmHg as long as infant is sensitive to CO₂ changes and pH remains > 7.25
- If peak pressures > 25 cmH₂O are needed to generate pre-operative tidal volumes, consider trial of decreased PEEP (as low as 3 cmH₂O) or transition to high-frequency oscillatory ventilation (HFOV)
- If there is evidence of a mass effect (e.g., mediastinal shift or decreased respiratory compliance), consider drainage and investigate etiology (e.g., tension pneumothorax, chylothorax), obtain chest X-ray and notify surgery

Nutrition

- Begin daily weights
- Higher caloric requirements are frequently required for up to 2 months post-operatively
- Utilize breastmilk (use donor human milk if maternal milk is not available)
- Regardless of gestational age (GA), advance feeds at the 32 - 34 weeks GA 1.5 - 2 kg Enteral Feeding Guideline pace ([ICN Enteral Feeding Guidelines](#))
- Fortify with hydrolyzed formula, (Alimentum/Pregestimil/Nutramigen)
 - For formula fed, transition to all hydrolyzed formula after demonstrating tolerance to full volume fortified donor milk
- Consult OT and engage Lactation as soon as enteral feeds are introduced

Post-Operative Pain Management

- Post-operative pain management should be individualized and guided by a clinically relevant and validated pain scoring tool
- Consider IV acetaminophen to reduce opioid requirements
- Monitor for neonatal withdrawal symptoms and wean accordingly

Ventilatory Weaning

- Due to increased need for sedation and risk of pulmonary hypertension, neonates are generally not weaned for 48 - 72 hours post-operatively
- **Earlier weaning may be necessary**
- Consider neurally-adjusted ventilatory assist (NAVA)
- Ensure extubation criteria are met

Earlier Ventilatory Weaning

- Over ventilation (CO₂ < 45)
- Over oxygenation (SpO₂ > 98%)
- Receiving large tidal volumes (> 6 mL/kg or more than received pre-operatively)

[CMH iNO Guidelines](#)

Before Discharge

- Record weight, length, and head circumference
- Obtain chest X-ray, echocardiogram, and brain MRI
- Ensure immunizations are up-to-date (including respiratory syncytial virus [RSV])
- Assess hearing
- Schedule outpatient follow up with NEON clinic, Cardiology, and Pediatric Surgery
- **Share discharge expectations, follow-up outpatient care, and long-term considerations with family**

Discharge Expectations

- Goal is breathing unassisted in room air if possible. If not possible, the goal is to maintain normal SpO₂ with a specified maximum flow rate
- 100% enteral feeds, ideally by mouth; however, CDH infants due to their prolonged respiratory course may need NG-tube (refer to [NICU Home NG Algorithm](#)) or G-tube

Follow-Up Outpatient Care

- Pediatric Surgery
- NEON clinic
- Cardiology

Long-Term Considerations

- Bowel obstruction
- Reflux
- Volvulus
- Recurrence
- Pulmonary hypertension
- Pulmonary hypoplasia

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Objective of Clinical Pathway

To provide care standards for neonates with suspected congenital diaphragmatic hernia or neonates and infants diagnosed with congenital diaphragmatic hernia. The Congenital Diaphragmatic Hernia Clinical Pathway guides the care process from the delivery room management for the neonate with suspected congenital diaphragmatic hernia through post-operative care following surgical repair to minimize care variation.

Background/Epidemiology

Congenital diaphragmatic hernia (CDH) can be described as a developmental closure defect occurring in approximately 1 of every 3600 live births in the United States, and up to 2% of infant mortalities (Kosiński & Wielgoś, 2017; Mai et al., 2019; Wynn et al., 2014). As the result of the diaphragmatic opening, abdominal organs herniate into the chest leading to pulmonary hypoplasia and pulmonary hypertension (Kosiński & Wielgoś, 2017; Wynn et al., 2014).

Prenatal imaging techniques, such as ultrasound, fetal echocardiography, and fetal magnetic resonance imaging, can be used to diagnose CDH, assist in determining its severity, and guide decisions for possible treatment options (Chandrasekharan et al., 2017). Thus, a multidisciplinary approach to antenatal management and postnatal management is essential to the care of the mother preparing for delivery and the neonate diagnosed with CDH (Chandrasekharan et al., 2017; Kosiński & Wielgoś, 2017; Sharma, 2017). The Congenital Diaphragmatic Hernia Clinical Pathway aims to provide a pragmatic multidisciplinary approach supported by evidence and expert opinion in the delivery room management for the neonate with suspected CDH through post-operative care following surgical repair when diagnosed.

Target Users

- Physicians (Neonatology, Pulmonology, Surgery, NICU Fellows, Residents, NICU Hospitalists)
- Nurse Practitioners
- Nurses (Delivery Room, NICU, Operating Room, ECMO)
- Respiratory Therapy
- Clinical Nutrition Specialist

Target Population

Inclusion Criteria

- Neonates with suspected CDH at the time of delivery
- Neonates and infants diagnosed with CDH

AGREE II

The Canadian Congenital Diaphragmatic Hernia Collaborative international guidelines provided guidance to the Congenital Diaphragmatic Hernia Clinical Pathway Committee (Puligandla et al., 2018, 2023). See Table 1 and Table 2 for AGREE II.

Table 1
AGREE II Summary for the Diagnosis and Management of Congenital Diaphragmatic Hernia: A Clinical Practice Guideline (Puligandla et al., 2018)

Domain	Percent Agreement	Percent Justification [^]
Scope and purpose	99%	The aim of the guideline, the clinical questions posed and target populations were identified.
Stakeholder involvement	88%	The guideline was developed by the appropriate stakeholders and represents the views of its intended users.
Rigor of development	94%	The process used to gather and synthesize the evidence, the methods to formulate the recommendations and to update the guidelines were explicitly stated.
Clarity and presentation	100%	The guideline recommendations are clear, unambiguous, and easily identified; in addition, different management options are presented.
Applicability	81%	Barriers and facilitators to implementation, strategies to improve utilization and resource implications were addressed in the guideline.

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Editorial independence	98%	The recommendations were not biased by competing interests.
Overall guideline assessment	93%	
See Practice Recommendations		

Note: Four EBP Scholars completed the AGREE II on this guideline.
 ^Percentage justification is an interpretation based on the Children’s Mercy EBP Department standards.

Table 2
 AGREE II Summary for the Diagnosis and Management of Congenital Diaphragmatic Hernia: A 2023 Update from the Canadian Congenital Diaphragmatic Hernia Collaborative (Puligandla et al., 2023)

Domain	Percent Agreement	Percent Justification [^]
Scope and purpose	100%	The aim of the guideline, the clinical questions posed and target populations were identified.
Stakeholder involvement	76%	The guideline was developed by the appropriate stakeholders and represents the views of its intended users.
Rigor of development	94%	The process used to gather and synthesize the evidence, the methods to formulate the recommendations and to update the guidelines were explicitly stated.
Clarity and presentation	99%	The guideline recommendations are clear, unambiguous, and easily identified; in addition, different management options are presented. Round nor are management options provided.
Applicability	76%	Barriers and facilitators to implementation, strategies to improve utilization and resource implications were addressed in the guideline.
Editorial independence	100%	The recommendations were not biased with competing interests.
Overall guideline assessment	91%	
See Practice Recommendations		

Note: Four EBP Scholars completed the AGREE II on this guideline.
 ^Percentage justification is an interpretation based on the Children’s Mercy EBP Department standards.

Practice Recommendations

Please refer to the Canadian Congenital Diaphragmatic Hernia Collaborative (Puligandla et al., 2018, 2023) clinical practice guidelines for diagnosis and management recommendations.

Additional Questions Posed by the Clinical Pathway Committee

No clinical questions were posed for this review.

Recommendation Specific for Children’s Mercy

Children’s Mercy adopted the majority of the practice recommendations made by the Canadian Congenital Diaphragmatic Hernia Collaborative Guidelines (Puligandla et al., 2018, 2023). Variations include:

- Regarding ventilation, partial pressure of carbon dioxide and pH levels slightly varied. The Canadian Congenital Diaphragmatic Hernia Collaborative Guidelines recommended pCO₂ between 45 and 60 mm Hg and pH between 7.25 and 7.40, whereas the Congenital Diaphragmatic Hernia Clinical Pathway Committee recommends pCO₂ between 45 and 65 mm Hg and pH between 7.20 and 7.40.

Measures

- Utilization of the Congenital Diaphragmatic Hernia Clinical Pathway

Value Implications

The following improvements may increase value by reducing healthcare costs and non-monetary costs (e.g., missed school/work, loss of wages, stress) for patients and families and reducing costs and resource utilization for healthcare facilities.

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- Decreased unwarranted variation in care

Organizational Barriers and Facilitators**Potential Barriers**

- Variability of acceptable level of risk among providers

Potential Facilitators

- Collaborative engagement across care continuum settings during clinical pathway development

Diversity/Equity/Inclusion

Our aim is to provide equitable care. These issues were discussed with the Committee, reviewed in the literature, and discussed prior to making any practice recommendations.

Power Plans

- The Congenital Diaphragmatic Hernia Clinical Pathway does not have an associated powerplan. However, the *ECMO for Congenital Diaphragmatic Hernia Repair Powerplan* can be utilized for neonates or infants when ECMO is indicated

Associated Policies

- Delivery Room Management of the Neonate with Congenital Diaphragmatic Hernia
- Initial Stabilization of the High-Risk Neonate in the Delivery Room
- Ex-Utero Intra-Partum Treatment (EXIT)

Clinical Pathway Preparation

This pathway was prepared by the Evidence Based Practice (EBP) Department in collaboration with the Congenital Diaphragmatic Hernia Clinical Pathway Committee composed of content experts at Children's Mercy Kansas City. If a conflict of interest is identified, the conflict will be disclosed next to the committee member's name.

Congenital Diaphragmatic Hernia Clinical Pathway Committee Members and Representation

- Ekta Patel, DO | Neonatology | Committee Chair
- Jennie Godwin, DO | Neonatology | Committee Member
- John Daniel IV, MD, MS | Neonatology, Neonatal ECMO Director | Committee Member
- Megan Gubichuk, MD | Pulmonology | Committee Member
- Richard Hendrickson, MD, FAAP, FACS | Pediatric Surgery | Committee Member
- Elise Loughman, MD | Neonatal-Perinatal Fellow | Committee Member
- Debra Newton, RN, MSN, CCRN | Director Extracorporeal Support, ECMO Program, Apheresis Program | Committee Member
- Mary Hagerty, APRN, NNP-BC | Neonatology | Committee Member
- Christina Elliott, RN-NIC | Core Charge Nurse – Intensive Care Nursery | Committee Member
- Lucy Pappas, MS, RD, CSPCC LD | Clinical Nutrition Specialist IV, Nutrition Services | Committee Member
- Amy Schnack, MS, MPH, RD, LD, IBCLC | Clinical Nutrition Specialist II, Nutrition Services | Reviewer

Patient/Family Committee Member

- Andrew Carr | Committee Member
- Barb Carr | Committee Member

EBP Committee Members

- Todd Glenski, MD, MSHA, FASA | Anesthesiology, Evidence Based Practice
- Kelli Ott, OTD, OTR/L | Evidence Based Practice

Clinical Pathway Development Funding

The development of this clinical pathway was underwritten by the following departments/divisions: Neonatology, Pulmonology, Pediatric Surgery, ECMO Program, Intensive Care Nursery, Nutrition Services, Patient and Family Engagement, and Evidence Based Practice.

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Conflict of Interest

The contributors to the Congenital Diaphragmatic Hernia Clinical Pathway have no conflicts of interest to disclose related to the subject matter or materials discussed.

Approval Process

- This pathway was reviewed and approved by the Congenital Diaphragmatic Hernia Clinical Pathway Committee, Content Expert Departments/Divisions, and the EBP Department; after which they were approved by the Division of Neonatology and the Medical Executive Committee.
- Products are reviewed and updated as necessary every 3 years within the EBP Department at CMKC. Content expert teams are involved with every review and update.

Review Requested

Department/Unit	Date Obtained
Neonatology	August 2024
Pulmonology	September 2024
Pediatric Surgery	August 2024
ECMO Program	August 2024
Intensive Care Nursery	August 2024
Nutrition Services	August 2024
Patient and Family Engagement	August 2024
Evidence Based Practice	July 2024

Version History

Date	Comments
September 2024	Version one (Delivery Room Resuscitation, Pre-Operative Management, Surgical Management, and Post-Operative Care algorithms and Congenital Diaphragmatic Hernia Clinical Pathway Synopsis developed)

Date for Next Review

- September 2027

Implementation & Follow-Up

- Once approved, the pathway was presented to appropriate care teams and implemented. Care measurements will be assessed and shared with appropriate care teams to determine if changes need to occur.
- Order sets/power plans consistent with recommendations were created or updated for each care setting
- Education was provided to all stakeholders:
 - Nursing units where the Congenital Diaphragmatic Hernia Clinical Pathway is used
 - Department of Neonatology, Pediatric Surgery, Pulmonology, and Nutrition Services
 - Providers from the Fetal Health Center and Intensive Care Nursery
 - Resident physicians
- Additional institution-wide announcements were made via email, hospital website, and relevant huddles.

Disclaimer

When evidence is lacking or inconclusive, options in care are provided in the supporting documents and the power plan(s) that accompany the clinical pathway.

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* These clinical pathways do not establish a standard of care to be followed in every case. It is recognized that each case is different, and those individuals involved in providing health care are expected to use their judgment in determining what is in the best interests of the patient based on the circumstances existing at the time. It is impossible to anticipate all possible situations that may exist and to prepare a clinical pathway for each. Accordingly, these clinical pathways should guide care with the understanding that departures from them may be required at times.