Congenital Diaphragmatic Hernia (CDH) Pathway v2.0: **Table of Contents**

Stop and **Review**

Inclusion Criteria

- All children with congenital diaphragmatic hernia post surgical repair who have met extubation criteria
- No major cardiac abnormalities
- Gestational age (GA): > 35 weeks

Exclusion Criteria

- GA <35 weeks
- Major cardiac comorbidities

CDH Care

Appendix

Last Updated: January 2023
Next Expected Review: April 2027

For questions concerning this pathway, contact:
CDHpathway@seattlechildrens.org

Congenital Diaphragmatic Hernia (CDH) Pathway v2.0: **NICU**

Stop and **Review**

Inclusion Criteria

- All children with congenital diaphragmatic hernia post surgical repair who have met extubation criteria
- Infants < 6 weeks
- No major cardiac abnormalities
- Gestational age (GA): > 35 weeks

Exclusion Criteria

- GA <35 weeks
- Major cardiac comorbidities
- Infants > 6 weeks

Follow CDH Summary guideline (*for SCH only*)

Extubate 6-8 cm H20 CPAP, minimum 30% FIO2

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Weaning Criteria

- Stable clinical exam – baseline or improving tachypnea, no tachycardia, no increase work of breathing
- PCO2 <50
- Serum bicarb <30
- FIO2 ≤ = 40%
- No or mild pulmonary hypertension
- Adequate weight/length gain as determined by team
- No evidence of increased gastroesophageal reflux disease (GERD)

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Once clinically stable able to transition to HFNC

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Occupational therapy (OT) consult when pt. is at 4L- OT to assess readiness for feeding/swallow evaluation
(See Nutrition and Feeding Therapy Guideline)

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Wean HFNC: Up to 2x week by 1 L/min

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Floor criteria reached 2L-4L HFNC, see policy: High Flow Nasal Cannula (HFNC),10200 (*for SCH only*)

Congenital Diaphragmatic Hernia (CDH) Pathway v2.0: **Floor**

Stop and **Review**

Inclusion Criteria

- All children with congenital diaphragmatic hernia post surgical repair out of ICU on HFNC
- No major cardiac abnormalities
- Gestational age (GA): > 35 weeks

Exclusion Criteria

- GA <35 weeks
- Major cardiac comorbidities

Weaning Criteria

- Stable clinical exam – baseline or improving tachypnea, no increase tachycardia, no increase work of breathing, tolerance of daily therapy, maintaining baseline oxygen saturations with no desaturations
- PCO2 <50
- Serum bicarb <30
- Adequate weight/length gain as determined by team
- Concurrent assessment with Nutrition and Occupational Therapy (Nutrition and Feeding Therapy Guideline)
- No evidence of increased GERD

Higher Risk Patients (any of the following)

- ECMO requirement
- Persistent pulmonary hypertension
- LHR <1.3
- Ventilator days > 30
- Prolonged non-invasive respiratory support 1-2 weeks
- Need for diuretic use

- Ensure pulmonary consult has been initiated
- Eligible to advance per sprinting schedule up to 2x weekly as tolerated
- Weaning no more then 1L per week
- Sprint to either RA or 0.25-0.5L NC based on presence of pulmonary hypertension or profound pulmonary hypoplasia
- **HFNC sprinting schedule begins at 2L:**
  - Begin with one 2 hour
  - Increase to two 2 hour
  - Increase to two 3 hour
  - Increase to one 6 hour
  - Increase to one 8 hour
  - Increase to one 12 hours total
  - Sprint off HFNC to either RA/LFNC for 24 hours

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Patients with pulmonary hypertension may need oxygen at discharge

Once off highflow for 3-7 days check labs. If labs, growth, and work of breathing are reassuring then baby can discharge.

Lower Risk Patients

- Primary repair
- No pulmonary HTN

- Eligible to wean support up to 3x weekly
- Wean off 2L HFNC to RA
- Pulmonary consult if unable to wean

Once off highflow for 1-2 days check labs. If labs, growth, and work of breathing are reassuring then baby can discharge.

See Discharge Phase

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Congenital Diaphragmatic Hernia (CDH) Pathway v2.0: **Nutrition and Feeding Therapy**

Stop and **Review**

Inclusion Criteria

- All children with congenital diaphragmatic hernia post surgical repair who have met extubation criteria
- No major cardiac abnormalities
- Gestational age (GA): > 35 weeks

Exclusion Criteria

- GA <35 weeks
- Major cardiac comorbidities

NICU

Once return of bowel function:

- Surgery and NICU teams to decide initiation of feeds
- Recommend starting 10 mL/kg/day divided in Q3 hour boluses feeds via NG tube
- Consider starting at omeprazole 0.7 mg/kg/day, increasing as needed with max of 1mg/kg/dose given twice daily for severe GERD
- Trophic feedings of less then 20mL/kg/day will run in addition to total fluid allowance
- Once feedings progress beyond 20 mL/kg/day, begin weaning parenteral nutrition and consider fortification
- If not tolerating bolus feeds, consider transition to continuous feeds
- If persistence symptoms of GERD and respiratory distress, consider post pyloric feeds

Transfer to Floor

- Verify total fluid (typically 120-140 mL/kg/day)
- Typically need to fortify to 24-28kcal/oz to meet estimated calorie & protein needs (120+ kcal/kg, 3+ g/kg protein)
  - 2 kcal/oz increase daily to expected goal
  - Nutrition to provide specific goals and or guidance
- Ensure on 1 mL/d Poly-vi-sol w/ iron (if on fortified breast milk)
- Consider additional iron supplementation for history of anemia
- Ensure occupational therapy is consulting—clinical feeding evaluation must be completed prior to initiation of oral feeds

Floor Management

- Assess weight & length trend, ensure goal weight gain
- Occupational therapy to assess infant feeding and readiness for oral feeding advancement. Discuss with general surgery and nutrition to balance with oxygen and enteral feeding weaning
- Assess for evidence of gastroesophageal reflux, consider adding wedge
- Consider Videofluoroscopic Swallow Study (VFSS) for patients with concerns on clinical swallow evaluation, evidence of aspiration on chest Xray, pulmonary hypertension that is not improving, worsening work of breathing or blood gases

Usually within 1-2 weeks of anticipated discharge

Feeding Regimen Planning

- Goal to transition from continuous feeds to bolus feeds. Criteria for transitioning to bolus feeds include stable laboratory work, no increase work of breathing, no clinical evidence of gastroesophageal reflux
- Start feeding compression by running bolus feeds over 2 hours and compress feeds as tolerated toward a goal of 30 minutes or gravity bolus
- If unable to tolerate 30 min bolus feeds or gravity bolus, then consider transition to daytime boluses only with a nighttime continuous infusion

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Next Expected Review: April 2027

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If you are a patient with questions contact your medical provider, Medical Disclaimer

Congenital Diaphragmatic Hernia (CDH) Pathway v2.0: **Discharge Criteria**

Stop and **Review**

Inclusion Criteria

- All children with congenital diaphragmatic hernia who are eligible for discharge
- No major cardiac abnormalities
- Gestational age (GA): > 35 weeks

Exclusion Criteria

- GA <35 weeks
- Major cardiac comorbidities

Required family teaching

- CDH
- Nasogastric (NG) Tube Feeding Instructions
- Nasogastric (NG) Tube Inserting Instructions
- Giving Medicine through a Nasogastric (NG) Feeding Tube
- Signs and symptoms of increased effort of breathing
- Car seat teaching
- Teach family feeding regimen, recipe for formula fortification, feeding advancement plan for home
- Feeding therapist to teach families safe oral feeding plan for home

Discharge Coordination

- Finalize home oral + tube feeding regimen
- Ensure discharge feedings prescriptions and home supplies are in place including supplements and multi vitamins
- Care Coordination to initiate homecare orders (feeding supplies, home monitors and oxygen as needed)
- Car seat challenge passed
- PCP identified and appointment scheduled within 3-7 days of discharge
- Referral to Birth to Three and Infant Developmental Follow up Clinic at University of Washington
- SCH follow-up appointments scheduled
- Vaccinations as indicated including Synagis

Please see the American Academy of Pediatrics for follow-up guidelines

Coordinated follow up after discharge

- High risk CDH patients should be seen by provider and nutritionist 1-2 weeks after discharge
- Lower risk CDH patients should be seen by provider and nutritionist 4 weeks after discharge
- Occupational Therapy (OT)/PT as indicated, in conjunction with CDH clinic follow-up

Coordinated 2-3 months post discharge

- General surgery
- Pulmonary
- Nutrition
- OT/PT if indicated
- Cardiology/Pulmonary Hypertension team if indicated
- Ensure follow-up with Infant Developmental Follow up Clinic at University of Washington by six months of age

Discharge Criteria

- Teaching complete
- Supplies ordered
- Follow up appointments scheduled
- Medically cleared by providers

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Summary of Version Changes

- **Version 1.0 (4/21/2022):** Go live.
- **Version 2.0 (1/23/2023):** The Nutrition and Feeding Therapy phase of the algorithm was modified at the recommendation of the pharmacy. Version 1.0 was approved by P&T on 11/15/2022 by expedite review.

Approval & Citation

Approved by the CSW Congenital Diaphragmatic Hernia Pathway team for April 21, 2022, go-live

This work is made possible by the contributions of the late Dr. Daniel Ledbetter. Dr. Ledbetter led efforts to write and standardize multidisciplinary protocols for infants with CDH. His expertise, care, and mentorship has contributed greatly to the outcomes of these children. We are honored to continue his mission while acknowledging no one will ever take his place.

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Retrieval Website: https://www.seattlechildrens.org/pdf/congenital-diaphragmatic-hernia-pathway.pdf

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Evidence Ratings

This pathway was developed through local consensus based on published evidence and expert opinion as part of Clinical Standard Work at Seattle Children's. Pathway teams include representatives from Medical, Subspecialty, and/or Surgical Services, Nursing, Pharmacy, Clinical Effectiveness, and other services as appropriate.

When possible, we used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial or cohort studies. The rating is then adjusted in the following manner (from: Guyatt G et al. J Clin Epidemiol. 2011;4:383-94, Hultcrantz M et al. J Clin Epidemiol. 2017;87:4-13.):

Quality ratings are *downgraded* if studies:

- Have serious limitations
- Have inconsistent results
- If evidence does not directly address clinical questions
- If estimates are imprecise OR
- If it is felt that there is substantial publication bias

Quality ratings are *upgraded* if it is felt that:

- The effect size is large
- If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
- If a dose-response gradient is evident

Certainty of Evidence

✪✪✪✪ High: The authors have a lot of confidence that the true effect is similar to the estimated effect
✪✪✪○ Moderate: The authors believe that the true effect is probably close to the estimated effect
✪✪○○ Low: The true effect might be markedly different from the estimated effect
✪○○○ Very low: The true effect is probably markedly different from the estimated effect
Guideline: Recommendation is from a published guideline that used methodology deemed acceptable by the team
Expert Opinion: Based on available evidence that does not meet GRADE criteria (for example, case-control studies)

Bibliography

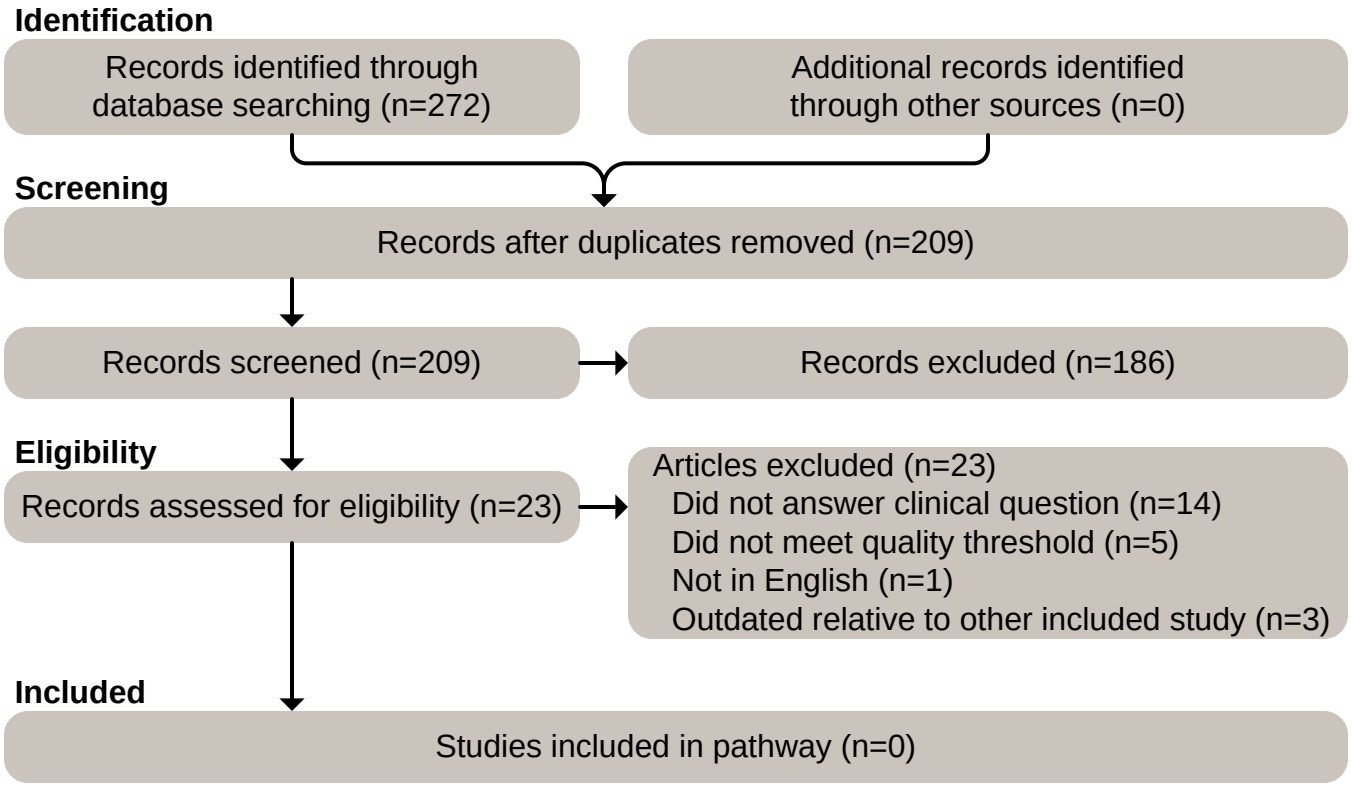
Literature Search Methods
A literature search was conducted May 2021 to target synthesized literature on congenital diaphragmatic hernias. The search was executed in Ovid Medline, Embase, and Turning Research into Practice (TRIP) databases. Results were limited to English language and items published January 2011 to May 2021.

Screening and data extraction were completed using DistillerSR (Evidence Partners, Ottawa, Canada). Two reviewers independently screened abstracts and included guidelines and systematic reviews that addressed optimal treatment, and prognosis of patients meet pathway inclusion/ exclusion criteria. One reviewer screened full text and extracted data and a second reviewer quality checked the results. Differences were resolved by consensus.

Literature Search Results
The searches of the 3 databases (see Electronic searches) retrieved 272 records. Our searches of other resources identified no additional studies that appeared to meet the inclusion criteria.

Once duplicates had been removed, we had a total of 209 records. We excluded 186 records based on titles and abstracts. We obtained the full text of the remaining 23 records and excluded 23.

We included no studies. The flow diagram summarizes the study selection process.



Flow diagram adapted from Moher D et al. BMJ 2009;339:bmj.b2535

Bibliography

Included Studies

None from structured search

Medical Disclaimer

Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required.

The authors have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication.

However, in view of the possibility of human error or changes in medical sciences, neither the authors nor Seattle Children's Healthcare System nor any other party who has been involved in the preparation or publication of this work warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such information.

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