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Chylothorax Management in the CVICU Clinical Pathway

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Rationale:

This clinical pathway was developed by a consensus group of Johns Hopkins All Children's Hospital (JHACH) physicians, advanced practice providers, nurses, and pharmacists to standardize the management of patients hospitalized in the Cardiovascular Intensive Care Unit (CVICU) with a diagnosis of chylothorax. It addresses the following clinical questions or problems:

1. When to evaluate for possible chylothorax
2. What studies are needed to confirm the diagnosis of chylothorax
3. Initial management strategies for chylothorax
4. Long-term treatment for chylothorax
5. When to adjust treatment strategy based on clinical response

Background:

Chylothorax is a fluid collection of fats and lymph fluid (chyle) within the pleural cavity. It typically results from leaking lymphatic vessels, with the most commonly affected vessel being the thoracic duct. This can be the result of a congenital lymphatic malformation, due to high venous pressures, or injury to the lymphatic system during congenital heart surgery. Numerous treatment options have been proposed for post-operative chylothorax, including intravenous (IV) medications, surgical ligation of the thoracic duct, and, more recently, interventional radiologic procedures to embolize the leaking lymphatic vessels, among others. Chylothorax can lead to significant morbidity, especially in the congenital heart disease (CHD) population, as it carries fat and fat-soluble vitamins, immunoglobulins, albumin, and often has a high protein content. Patients with chylothorax can also lose a significant amount of antithrombin III (ATIII), which can put a patient at risk of thrombosis, especially in a patient population with frequent need for antiplatelet therapies.[1-3]

Diagnosis:

Due to the large triglyceride component in chylous fluid, it tends to have a milky appearance. However, many post-operative patients have not started consuming a diet with a significant fat content; therefore, the fluid may continue to appear serous. Several characteristics of chylous fluid raise concern for chylothorax, including:

- Milky appearing fluid draining from a pleural drain
- Total chest tube output (CTO) > 20 mL/kg/day at > 24 hours post-operation
- Significant increase in fluid output after a patient begins consuming a full-fat diet

Several tests can help in diagnosing chylothorax when concerns are raised about the quality or quantity of the pleural fluid draining from the patient's chest tubes:

Pleural fluid diagnostic studies:

- Triglycerides (> 100 mg/dL)
- Cell count and differential with lymphocyte predominance (> 80%)
- Positive chylomicrons
- Pleural > serum triglycerides

Radiologic diagnostic studies:

- Head/neck ultrasound (US)
- Echocardiogram (ECHO) to evaluate for superior vena cava (SVC) thrombus (at provider discretion)
- Perioperative risk factors: previous chylothorax, elevated venous pressure, right heart dysfunction, venous thromboembolism (VTE), Fontan physiology, repeat sternotomy

An interdisciplinary group developed the following flowchart to help standardize care of chylothorax in the post-operative cardiac critical care population. The boxes listed after the flowchart help to guide decision-making and testing to appropriately detect, evaluate, and treat chylothorax in this population.

Clinical Management:

While interventional or surgical options can be considered, medical and pharmacologic interventions remain the initial mainstay of treatment across institutions.

Table 1: Diet modifications following chylous effusion diagnosis (See Box 3 for complete instructions on feeding and fortification)

Age	Formula Type	Options
< 1 year of age	Defatted human milk	• Fortify as below with a medium-chain triglyceride (MCT)-based formula for < 1 year of age • Supplement with vitamins A, D, E, and K • Supplement with walnut oil for essential fatty acids (EFAs)
	MCT-based enteral formulas	• Enfaport™ Supplement with vitamin D 400 units/day • Supplement with walnut oil for EFAs
1 – 13 years of age	MCT-based enteral formulas	• Monogen® • Milk protein intolerance – Tolerex® or Vivonex® Pediatric[1] • Supplement with walnut oil for EFAs
[1]Vivonex® Pediatric does not require MCT or walnut oil supplementation, but Tolerex® requires both MCT and walnut oil supplementation MCT and EFA-based calorie modulars: • MCT oil (8 calories/mL), Nutricia Liquigen® (4.5 calories/mL) • EFA modular: walnut oil (8 calories/mL)		

Table 2: Secondary pharmacologic interventions (See Box 4 for complete list)

Medication	Dose	Considerations
IVIG	400 mg/kg/dose IV	Consider if serum IgG < 400 mg/dL or < 600 mg/dL for patients with acute infection/immunosuppression
ATIII	Per the equation in Box 4	Limit to 2x/week Consider replacement for serum activity level < 60 – 80%, especially if requiring therapeutic anticoagulation
FFP	Minimum 10 mL/kg	Alternative to ATIII considering lower relative cost
Somatostatin analogue (octreotide)	40 mg/kg/day divided q8h SQ **OR** 3 mcg/kg/hour IV infusion	Consider after 2 – 6 days of high-volume output
Alpha-1 agonist (midodrine)	0.1 mg/kg/dose q8h enterally	Lymphatic vasoconstriction [Liou 2013]
Phosphodiesterase-5 inhibitor (sildenafil)	2 – 4 mg/kg/day divided q8h enterally	Inhibits lymphatic endothelial cell proliferation [Malleske, Yoder, 2015; Danial 2014; Wang 2017]
Non-selective beta-blocker (propranolol)	0.5 – 2 mg/kg/day divided q8h enterally	Vascular endothelial growth factor inhibitor [Liviskie 2020]
Systemic corticosteroids	Indication, timing, and dosing not specific [Loomba 2021-PHIS data]	Dexamethasone and methylprednisolone were associated with decreased LOS and secondary interventions in postoperative chylothorax patients Hydrocortisone is associated with decreased LOS [case studies – Fakhri, 2019; Fumihiro Miura, 2006]

Abbreviations: FFP, fresh frozen plasma; IgG, immunoglobulin G; IVIG, Intravenous immunoglobulin; LOS, length of stay; SQ, subcutaneous

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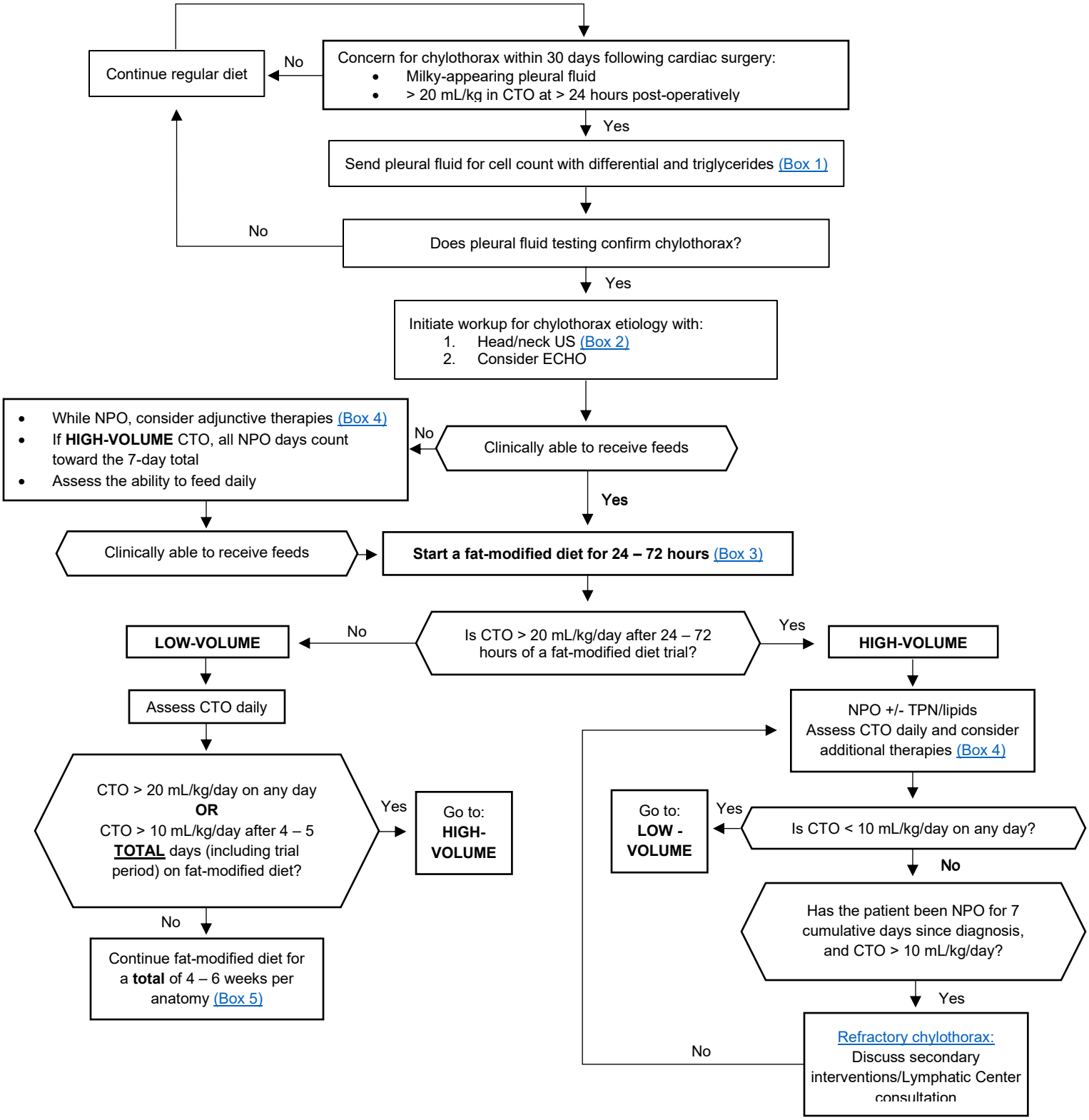

Abbreviations: TPN, total parenteral nutrition

Box 1: Diagnostic studies

Concern for chylothorax may include:

- Milky CTO
- CTO > 20 mL/kg beginning 24 hours after chest closure
- Surgeon's concern based on surgical findings, etc.

Pleural fluid studies confirmatory for chylothorax: (at least 1 of the following must be positive)

- **Triglycerides (> 110 mg/dL)**
- **Cell count with differential showing lymphocyte predominance (> 80%)**
- Positive chylomicrons
- Pleural > serum triglycerides

Box 2: Etiology work-up

Venous Doppler head/neck US:

- If VTE, anticoagulation per the *Management of Anticoagulation Therapy* policy

ECHO to include evaluation of SVC for thrombus:

- Peri-operative risk factors include:
  - Previous chylothorax
  - Elevated venous pressure
  - Right heart dysfunction
  - VTE
  - Fontan physiology
  - Repeat sternotomy

Box 3: Diet modifications following chylous effusion diagnosis

Defatted human milk:

- Fortify as below with an MCT-based formula for patients < 1 year
- Supplement with vitamins A, D, E, and K

MCT-based enteral formulas:

- < 1 year: Enfaport™ (83% MCT) Supplement with vitamin D 400 units/day
- 1 – 13 years: Monogen®* (84% MCT), Vivonex® Pediatric* (for protein milk allergy, 70% MCT), or Tolerex®** (for milk protein allergy, fat-free)

**Off-label use in patients < 1 year, however, nutritionally complete*
***Off-label use in patients < 3 years, needs MCT and EFA supplementation*

MCT and EFA-based calorie modulars:

- MCT oil (8 calories/mL), Nutricia Liquigen® (4.5 calories/mL)
- EFA modular: walnut oil (8 calories/mL)

Oral fat-modified diet:

- Start ≤ 10 grams fat from long-chain triglycerides to provide 2 – 4% total calories from linoleic acid (LA) and 0.25 – 0.5% from alpha linoleic acid (ALA)

Box 3: Diet modifications following chylous effusion diagnosis (continued)

EFA deficiency monitoring:

- EFA deficiency may develop in 1 – 3 weeks on no/low exogenous EFA delivery
    - Watch for physical symptoms including:
        - Dry/scaly rash
        - Sparse hair
        - Poor wound healing
    - For prolonged restriction or manifestation of symptoms:
        - Obtain EFA profiles
        - Monitoring triene to tetraene ratio (> 0.4 diagnostic of EFA deficiency)
        - Monitor liver function tests (LFTs)
        - Monitor platelets

Parenteral nutrition:

- Optimize calories
- Increase to goal protein:
    - Preterm: 3.5 – 4 g/kg
    - 0 – 2 years: 3 – 4 g/kg
- Lipids per needs/tolerance*: Minimum 0.5 g/kg Intralipid® (IL) or 2.5 g/kg SMOFlipid® for < 1 year of age or 2 g/kg SMOFlipid® for > 1 year of age to deliver ~ 0.1 g/kg LA
- For additional guidance, see the *Guidelines for Parenteral Nutrition* policy
- Screening labs:
    - Daily: Complete metabolic panel (CMP), magnesium, and phosphorus, until stable
    - 1 – 2 times per week: triglycerides

Box 4: Additional therapies

Refractory chylothorax:

Following diagnosis of chylothorax, a patient requiring 7 cumulative days of NPO with CTO remaining > 10 mL/kg/day qualifies for discussions regarding refractory chylothorax and secondary interventions. Discuss next steps and dosing of medications.

Work-up before Lymphatic Center referral:

Complete ECHO and diagnostic cardiac catheterization are recommended

IVIG[1]:

- Recommended dose: 400 mg/kg/dose IV
- Consider replacement if serum IgG < 400 mg/dL or < 600 mg/dL for patients with acute infection/immunosuppression
- Side effects: systemic vasodilation, transfusion reaction, anaphylactoid reaction

[1]*Consider pre-treatment with acetaminophen and diphenhydramine*

Box 4: Additional therapies (continued)

ATIII:

- Recommended dose: [(120 – patient's ATIII) x patient's weight (kg)] / 1.4
- Limit replacements to 2x per week
- Consider replacement for serum ATIII activity level < 60 – 80% (especially if requiring therapeutic anticoagulation)

FFP:

- As an alternative, in consideration of the cost of ATIII

Volume replacement recommendations:

- Consider maintaining serum albumin > 3 mg/dL with albumin 25% 0.5 – 1 g/kg/dose q6h IV for 24 hours
- Caution using additional volume during the early postoperative period
- Consider utilizing/alternating products for replacement: FFP, albumin 5%, and/or Lactated Ringer's (LR)

Consider venogram with high-volume chylothorax, regardless of the results of US

Somatostatin analogue (octreotide):

- Recommended dose: 40 mg/kg/day divided q8h SQ OR 3 mcg/kg/hour IV infusion
- Consider after 2 – 6 days of high-volume output
- Aggressive **diuretic** management tailored to the patient's clinical status

Alpha-1 agonist (midodrine) – lymphatic vasoconstriction:

- Recommended dose: 0.1 mg/kg/dose q8h enterally [Liou, 2013]

Phosphodiesterase-5 inhibitor (sildenafil) – inhibits lymphatic endothelial cell proliferation:

- Recommended dose: 2 – 4 mg/kg/day divided q8h enterally [Malleske, Yoder 2015; Wang 2017]

Non-selective beta-blocker (propranolol) – vascular endothelial growth factor inhibitor:

- Recommended dose: 0.5 – 2 mg/kg/day divided q8h enterally [Liviskie 2020]

Systemic corticosteroids:

- Dexamethasone and methylprednisolone were associated with decreased LOS and secondary interventions in postoperative chylothorax patients. Hydrocortisone was associated with decreased LOS. [Indication, timing, and dosing not specified; Loomba, 2021 - PHIS data] [Case studies - Fakhri, 2019; Fumihiro Miura, 2006]

Box 5: Fat-modified diet duration

Literature demonstrates a traditional fat-modified diet ranging from 2 to 8 weeks

Duration of therapy for patients with superior caval anastomoses is 6 weeks, and 4 weeks for all other postoperative patients

References:

1. Krishnamurthy, M.B/, & Malhotra, A. (2017). Congenital chylothorax: current perspectives and trends. *Research and Reports in Neonatology*, 53–63. https://doi.org/10.2147/rrn.s128703
2. Bernet-Buettiker, V., Waldvogel, K., Cannizzaro, V., & Albisetti, M. (2006). Antithrombin activity in children with chylothorax. *European Journal of Cardio-Thoracic Surgery*, *3*, 406–409. https://doi.org/10.1016/j.ejcts.2005.12.015
3. Tutor, J. D. (2014). Chylothorax in Infants and Children. *Pediatrics*, *4*, 722–733. https://doi.org/10.1542/peds.2013-2072

Outcome Measures:

- Chest tube duration
- Chest tube replacement
- Chylothorax recurrence
- LOS
- Patient adherence to a low-fat diet

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