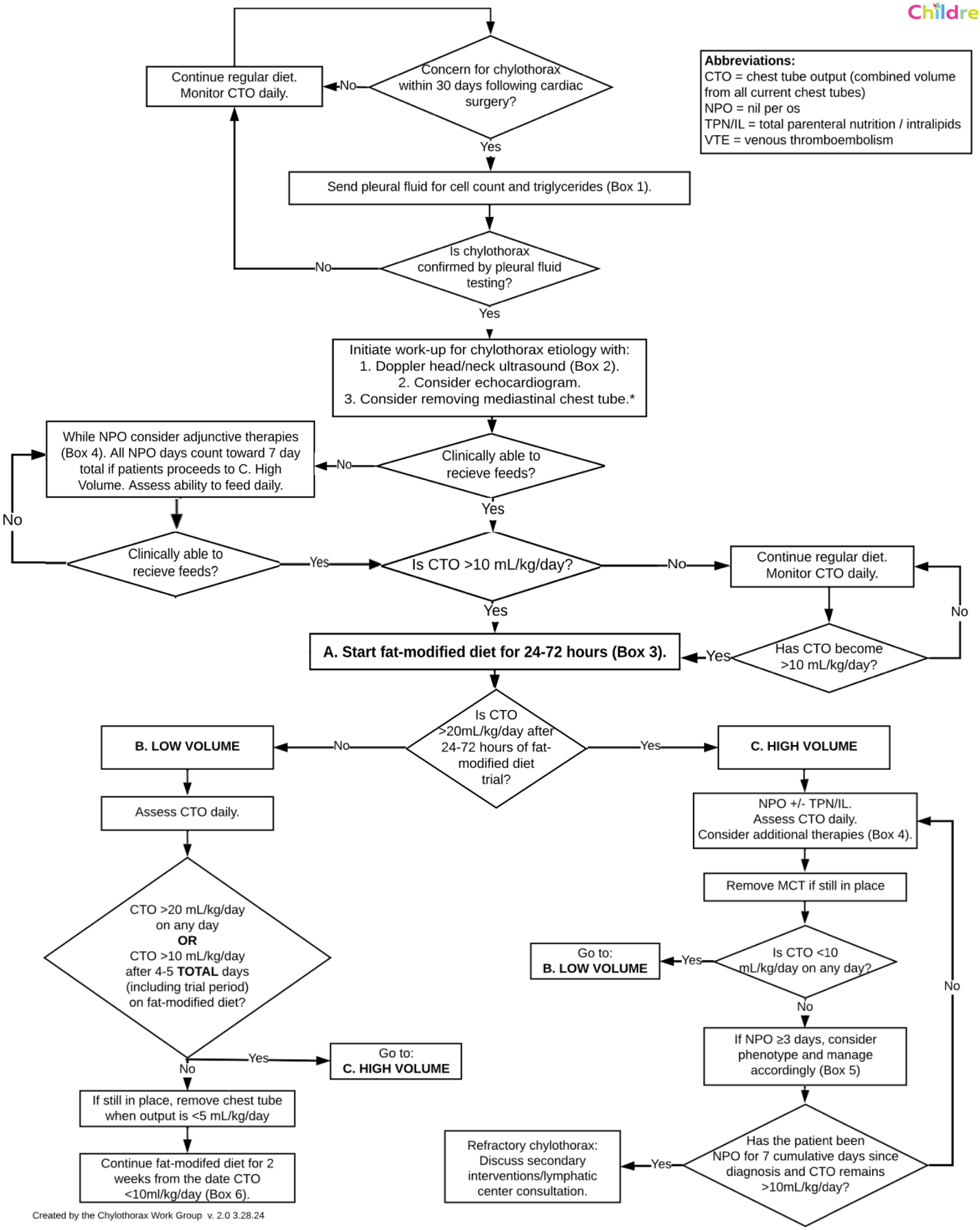
Diagnosis & Management of Chylothorax After Pediatric Cardiac Surgery



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Introduction: Chylothorax following pediatric cardiac surgery is a known complication associated with significant morbidity, mortality, and cost and can be compounded by significant variations in management, including dietary modifications and surgical interventions. UNC has partnered with the first multi-institution chylothorax work group through PC4 and has adopted a multi-institution algorithm to standardize care and management of chylothorax following cardiac surgical repair with smart aims of reducing median chest tube duration and prescribed length of dietary modifications.

BOX 1: Diagnostic Studies to Consider

Concern for chylothorax may include: milky appearing CTO, CTO >20mL/kg/day beginning 24 hrs after chest closure, surgeon 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) - *"Triglycerides, body fluid" in EPIC*
- ***Lymphocyte predominance*** (>80%) - *"Body fluid cell count" in EPIC*
- Pleural triglycerides > serum triglycerides
- Positive chylomicrons
    - Consider chylomicron testing if definitive diagnosis required, used to distinguish between chylous and nonchylous effusions
    - Tests are not mapped in EPIC – must be requested as "referral lab test, other" and are send out labs to Mayo Clinic
        - Lipoprotein Metabolism Profile (serum) – Test ID: LMPP
            - https://www.mayocliniclabs.com/test-catalog/overview/83673
            - Performed Mon-Th & Sat, turn-around time of 2-4 days (from time sample received)
        - Lipid Analysis (body fluid) – Test ID: BFLA1
            - https://www.mayocliniclabs.com/test-catalog/overview/614164
            - Performed Mon-Fri, turn-around time of 3-4 days (from time sample received)

BOX 2: Etiology Work-up (Risk factors: previous chylothorax, elevated venous pressure, decreased function, and VTE)

- Venous doppler head/neck ultrasound: If VTE present, provide anticoagulation per institutional standard.
- Echo to include evaluation of SVC for thrombus and consider venogram with negative echo but suspicion for VTE remains high.

**Contraindications to removal of mediastinal chest tube: high risk for bleeding or tamponade, bloody chest tube output, open chest. Replace with pleural chest tube if clinically indicated.*

BOX 3: Diet Modifications Following Chylous Effusion Diagnosis

Defatted human milk, fortify as below with MCT based formula for <1 year of age. Supplement with ADEK multivitamin.

MCT based enteral formulas:

< 1 year of age: Monogen*, Enfaport, Lipstart*, or Vivonex* (for milk protein intolerance). Supplement with 400 IU Vitamin D/day.

1-13 years of age: Monogen, Vivonex Pediatric (for milk protein intolerance), or Portagen**

Medium Chain Triglyceride (MCT) Based Calorie Modulars: MCT oil (8 Cal/ml), Nutricia Liquigen (4.5 Cal/ml), Vitaflo MCT Procal (112 Cal/1 powder packet).

Oral Fat Modified Diet:

Start ≤10g fat/day from long-chain triglycerides to provide 2-4% total calories from linoleic acid (LA) and 0.25-0.5% from alpha-linolenic acid (ALA).

Essential Fatty Acid (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:tetraene ratio (>0.4 diagnostic of EFA deficiency), LFTs, platelets.

Parenteral Nutrition: Optimize calories and protein. Protein may need to be increased by 1.5-2x RDA/DRI (0-2 years old: provide 3-4 g/kg; preterm: provide 3.5-4g/kg).

Dose lipids (IL) per needs/tolerance but with minimum of 0.5g/kg/day 20% IL or 1g/kg/day SMOF (goal is 2g/kg/day SMOF for 0-12 months of age) in order to deliver ~0.1g/kg/day LA. For preterm infants, double lipid dose in order to deliver the minimum 0.25g/kg/day LA to prevent EFA deficiency.

Parental Nutrition Screening Labs*:* daily until stability: electrolytes (BMP) including Mg, PO4; 1-2x/week: triglycerides, LFTs and albumin.

*Nutritionally complete but off label use in patients less than 1 yo

**Not nutritionally complete, requires trace mineral supplementation

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BOX 4: Additional Therapies for consideration in high output chylothorax

Replacement therapies:

- ***Intravenous Immunoglobulin*:** Recommended dose: 400 mg/kg IV. Replacement for serum level <400 (consider replacement for level <600 with acute infection or immunosuppressed pt). Limit replacement to twice/week. Consider pre-treatment with Tylenol and Benadryl.
- ***Antithrombin III*:** Recommended dose: 500 Units=1 vial for serum activity level <60-80%, especially if requiring therapeutic anticoagulation or at higher risk for thrombosis (i.e. shunted patients, nephrotic patients, etc.). Limit replacement to twice/week. Consider FFP as an alternative to ATIII and for volume replacement.
- ***25% Albumin*:** Recommended dose 0.5-1g/kg IV q6h x24h to keep serum Albumin >3.0 mg/dl.
- ***Volume replacement*:** Consider alternating the following products for replacement: FFP, 5% albumin, and/or LR. Caution using additional volume during early postoperative period.

Adjunctive therapies:

- ***Somatostatin Analogue* (Octreotide):** Recommended dose: 3 mcg/kg/hr IV gtt (may double dose daily to max 10mcg/kg/hr if no effect).
- ***Alpha-1 Agonist / lymphatic vasoconstriction* (Midodrine):** Recommended dose: 0.1 mg/kg PO q8h
- ***Phosphodiesterase-5 inhibitor* (Sildenafil):** Recommended dose: 2-4 mg/kg/day PO divided q8h.
- ***Non-selective beta-blocker / vascular endothelial growth factor inhibitor* (Propranolol):** Recommended dose: 0.5-2 mg/kg/day PO divided TID
- ***Systemic corticosteroids*:** dexamethasone, methylprednisolone or hydrocortisone

Prior to referral to lymphatic center, complete echocardiogram and diagnostic cardiac catheterization are recommended.

BOX 5: Phenotyping

Mechanical disruption/Lymphatic compression

Presentation: Surgeon suspicion for direct injury; vascular ring repair. Consider lymphatic compression (e.g. chest tubes have been reported to compress lymphatic structures)

Management: Discuss early (before POD 7) thoracic duct ligation for suspected direct injury or evaluation for lymphatic compression

Venous hypertension

Presentation: persistently elevated CVP, RV non-compliance on echo, significant LA hypertension, obstructed venous flow (i.e. thrombus), long term IJ CVL (>7 days), small distal pulmonary arteries in Glenn physiology, elevated mean airway pressure

Studies to consider: complete echo, upper/lower extremity ultrasounds with doppler, venogram to eval for external venous compression, MR lymphangiogram

Management: investigate for residual cardiac lesions, increase milrinone, decrease mean airway pressure or extubate if able, treat clot if present, consider prophylactic anticoagulation if high risk for clot, aggressive diuresis (negative fluid balance), consider discussing case with a lymphatic center

Pro-inflammatory

Presentation: profound capillary leak, fluid non-responsive, inability to wean vasopressors (remains on 2+ pressors), inability to achieve negative fluid balance, elevated BUN, vasoplegia with low CVP

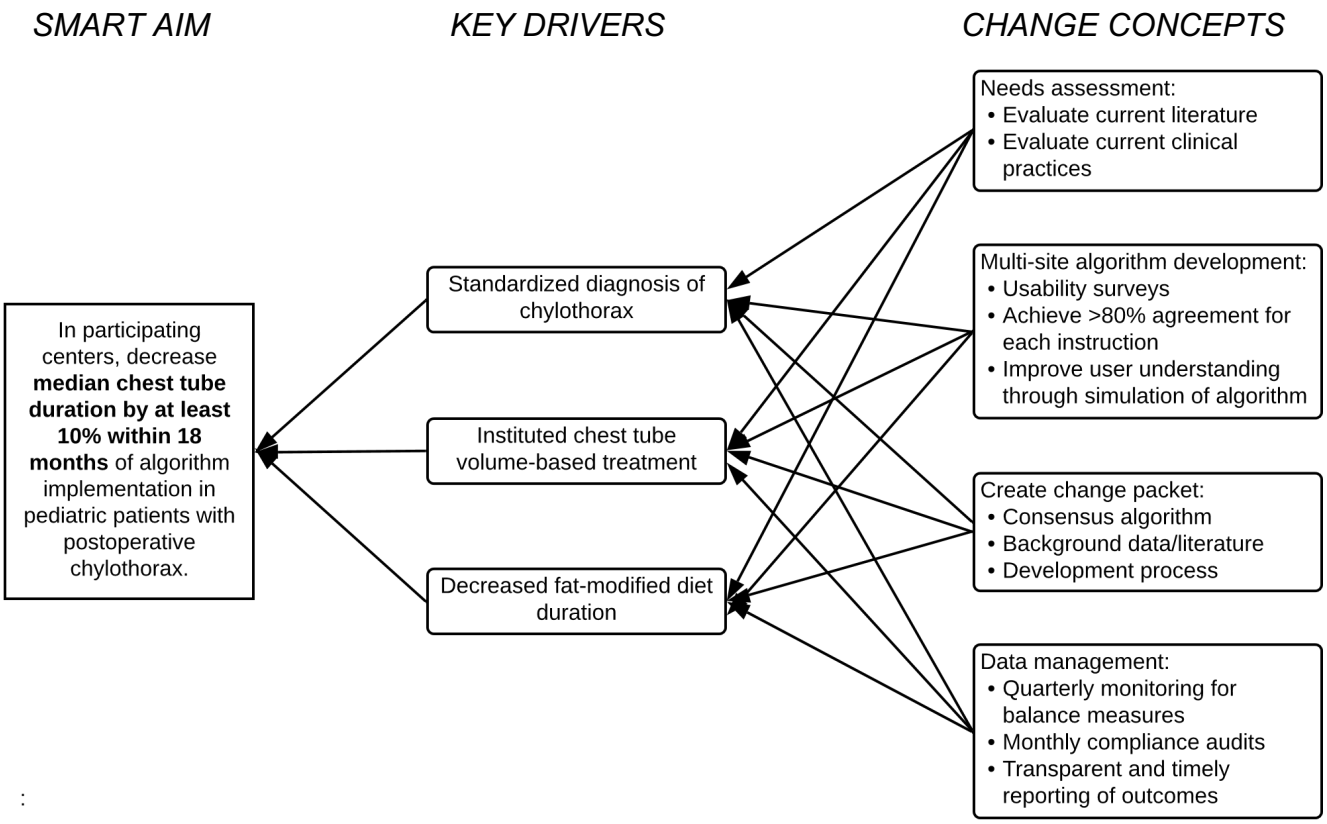
Studies to consider: serum inflammatory markers, serum ferritin, infectious work-up

Management: treat any infection, methylprednisolone for 5 days, vasoactives as needed, consider rheumatology consult

BOX 6: Fat-modified Diet Duration

Literature demonstrates traditional fat-modified diet ranging from 2-8 weeks. Current experience within the Chylothorax Work Group demonstrates the efficacy of a 2-week fat-modified diet duration in preventing recurrence of chylothorax (Winder MM, Schwartz S, Buckley JR, et al. Optimal Fat-Modified Diet Duration for the Treatment of Postoperative Chylothorax in Children. **Ann Thorac Surg**. Published online June 10, 2023).

Based on current institutional standards, **UNC will utilize a 2-week duration of fat-modified diet.**



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Process aim >=80% compliance at participating centers
Compliance = each instruction is followed within 48 hours of prescribed timing.

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