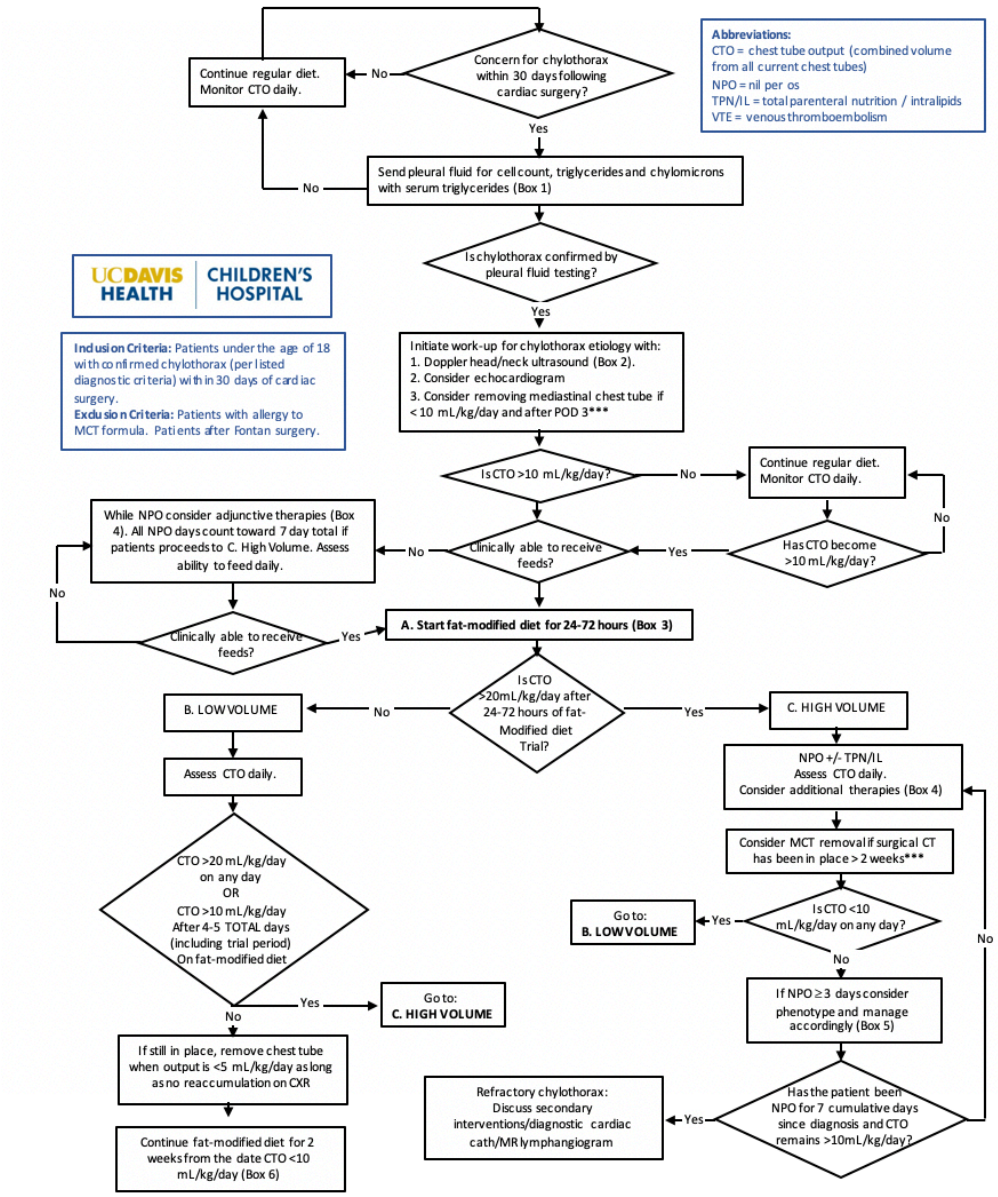
UCDAVIS HEALTH | CHILDREN'S HOSPITAL

Diagnosis and Management of Chylothorax after Pediatric Cardiac Surgery



BOX 1: Diagnostic Studies to Consider

Concern for chylothorax may include: milky CTO, CTO >20mL/kg/day beginning 24 hours 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)
- Lymphocyte predominance (>80%)
- Positive chylomicrons
- Pleural>serum triglycerides

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 chest tube with pleural tube if clinically indicated.*

Box 3: Diet Modifications Following Chylous Effusion Diagnosis

MCT based enteral formulas:

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

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

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 to 1.5-2x RDA/DRI. (Ex. 0-2 year olds: 2.5-4 g/kg, preterm: 3-4 g/kg).

Intravenous lipid emulsions are a safe and effective method of providing EFA for patients with chylothorax. Lipids are delivered directly into the bloodstream, bypassing the lymphatic system. Minimum dose of 0.5 g/kg/day Intralipids 20% or 1.5 g/kg/day SMOF lipid 20% is required. For preterm infants, a minimum dose of 1 g/kg/day Intralipids 20% or 2.5 g/kg SMOF lipid 20% is needed to deliver minimum of 0.25 g/kg/day LA to prevent EFA deficiency.

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

*Not nutritionally complete for infants - if remain on Monogen for longer than 2 weeks, please check essential fatty acid panel and add LCT supplementation

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

BOX 4: Additional Therapies for consideration in high output chylothorax

Replacement therapies: Recommend sending albumin, coagulation factors and IgG levels q48 – 72 hours

·**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:

·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

·Somatostatin Analogue (Octreotide): Recommended dose: 40 mcg/kg/day SQ divided q8h or 3mcg/kg/hr IV gtt (may double dose daily to max 10mcg/kg/hr if no effect) – only consider if above adjunctive therapies completed/contraindicated or secondary lymphatic center recommends

Prior to referral to lymphatic center, complete echocardiogram, diagnostic cardiac catheterization and MR lymphangiogram should be completed. Prior to MR lymphangiogram please obtain abdominal US.

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).

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