Chylothorax Algorithm

Introduction
Chylothorax refers to the accumulation of lymphatic fluid in the pleural space. Chylothorax following cardiac surgery is the most common cause of chylothorax in pediatrics and occurs because of direct intraoperative thoracic duct injury, central venous thrombosis, or elevated central venous pressure. Postoperative chylothorax following repair for congenital heart surgery is a complication associated with significant morbidity and mortality and treatment options are challenging given the nature of the diagnosis. The development of a clinical practice guideline for management of chylothorax following cardiac surgical repair has been noted to provide earlier diagnosis, reduced ICU and total hospital length of stay, decreased days of mechanical ventilation, and shorter chest tube duration. 3,5,9,11

Diagnosis
- Chylothorax should be suspected if output is turbid or “milky” in appearance or with high output 7
- High risk patient populations include: 8,9,10
  - Heart transplant
  - Cavopulmonary anastomoses (Glenn & Fontan)
  - Tetralogy of Fallot
  - Transposition of the great vessels
  - Aortic arch anomalies
  - TAPVR
- Pleural fluid should be sent for the following studies [orders written as appears in EPIC]:
  - Aerobic/Anaerobic culture (only with NEW sterile insertion of chest tube)
  - Body Fluid Cell Count
  - Triglycerides, Body Fluid
  - Protein, Body Fluid
  - Lactate Dehydrogenase, Body Fluid
  - Albumin Level, Body Fluid
- Diagnosis of chylothorax is established with:
  - Pleural fluid with:
    - Triglyceride level > 110 mg/dL, WBC > 1,000 cells/ul, Lymphocyte fraction > 80% 4,5,9
    - Note that triglyceride level may be below diagnostic threshold in fasting patients, therefore essential to assess for lymphocyte predominance
    - OR if the ratio of pleural fluid triglycerides : serum triglycerides is > 1
- Chylothorax can be classified as either high or low output: 11
  - Low output: CT output <= 20 mL/kg/day
  - High output: CT output > 20 mL/kg/day
- Chylomicron Testing
  - If definitive diagnosis required, used to distinguish between chylosus and nonchylosus effusions
  - Note that these tests are not currently mapped in EPIC therefore they have to be requested as “referral lab test, other” and are send out labs to Mayo Clinic
  - Test Options:
    - 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)

Laboratory Evaluation after Diagnosis
- Consider echocardiogram to assess for chylopericardium and elevated central venous pressure
- Consider PVLs to assess for thrombus (if upper extremity / neck line present)
If thrombus present, consider anticoagulation therapy and hematology consult

- At diagnosis of chylothorax, send: Albumin level, DIC Profile, and IgG level
  o Consider replacement with 25% (1 g/kg x 4 doses scheduled 6 hours apart) if albumin level if < 3.0
  o Consider replacement with appropriate factors for coagulopathy
  o Consider replacement with IVIG 1 g/kg for IgG level < 200
- Send Albumin, AT3, IgG, and DIC profile at least MWF for high output chylothorax and weekly for low output

**Management Strategies**

- Limiting the duration of chylous loss and optimizing nutrition is key to prevent complications of chylothorax including micronutrient deficiencies, malnutrition, immunologic losses, sepsis, and poor wound healing\(^1\)
- Consider drainage of pleural fluid via thoracostomy if pleural chest tubes are not currently in place\(^5,7\)

**Dietary Considerations**

- First line management of low output chylous effusion includes dietary modifications to a low-fat, high medium chain triglyceride (MCT) formula\(^7\)
  ▪ Formulary options include Monogen or Enfaport
- First line management of high output chylous effusion includes total NPO status with TPN / IL
  ▪ Consider transitioning to oral/enteral nutrition with MCT enriched diet after 7 days if CT output <10mL/kg/day
  ▪ Consider fat soluble vitamin supplementation if chylous output remains high output > 1 week
  ▪ IV fat emulsion may be required to supplement oral/enteral nutrition for calorie and fat intake
  ▪ After 4 weeks on low-fat diet, consider sending essential fatty acid profile (may require supplementation to avoid deficiency)
- Include consultation with a Registered Dietitian to guide nutrition decisions

**Medication Considerations**

- Octreotide [Standostatin]
  o Consider initiating infusion for refractory high output chylothorax if CT output is not <10 mL/kg/day after maintaining NPO status with TPN for 7 days\(^3,5\)
    ▪ Recommended laboratory monitoring includes LFTs, TFTs, and blood glucose
    ▪ Contraindications include vascular compromise\(^9\)
    ▪ Note that there is higher risk of NEC when used in neonatal populations\(^5\)
  o Initiate continuous IV infusion beginning at 2 mcg/kg/hr, increase by 2 mcg/kg/hr q24h up to a maximum of 10mcg/kg/hr
  o Continue infusion for a total of 7 days, then begin slow wean by 1 mcg/kg/hr q24h
  o If at 7 days CT output is <10 mL/kg/day, initiate enteral feeding with chylothorax diet
  o If at 7 days CT output remains >10 mL/kg/day, consider next interventions
  o Treatment effect typically seen at 5-6 days, with median reported at 10-18 days
  o Infusion should be gradually weaned by 1 mcg/kg/day
- Fungal Prophylaxis
  o Fungal prophylaxis may be indicated for patients who are NPO with TPN > 7 days with another major or minor risk factor
  o Recommended starting antifungal is Fluconazole unless contraindicated
- Prednisone
  o Consider prednisone (1mg/kg/day divided BID for 5-7 days) for patients with refractory chylothorax

**Interventions for Refractory Chylothorax**

- Consider MRI and lymphangiography to determine the etiology of chylothorax\(^10\)
- Chemical Pleurodesis with Doxycycline\(^5\)
  o Consider in patients who are refractory to NPO status and octreotide therapy
  o Typical dose: 20 mg/kg instilled into intrapleural space via pleural chest tube, dwell for 6 hours while repositioning patient every 30 minutes as tolerated, then open chest tube to drain
Give with concurrent intrapleural lidocaine for analgesia (Lidocaine 1% PF 1.5 mg/kg, dispensed as vial)

- Provide adequate patient-specific sedation and analgesia, as procedure can be very painful

Surgical Interventions

- Includes mechanical pleurodesis / thoracic duct ligation / pleuroperitoneal shunt
- Surgical therapy is typically reserved for cases with persistent output after 4 weeks

Chest Tube Removal

- Chest tube removal algorithms can significantly reduce duration of chest tubes and overall ICU and hospital lengths of stays

- General Guidelines for all Postop Patients:
  - Consider chest tube removal if volume over the last twelve hours is:
    - < 10 mL for < 10 kg
    - < 40 mL for 10-20 kg
    - < 60 mL for 20-40 kg
    - < twice body weight for > 40 kg
  - Defer removal and re-evaluate the next day if effusion or pneumothorax present on CXR

- For patients with history of chylothorax
  - Wait until enteral nutrition advances > 50% of goal prior to considering CT removal, then refer to the above criteria

Disclaimer

(1) This pathway is a general guideline and does not represent a professional care standard. Care should be revised and individualized to meet individual patient needs.

(2) This is a quality improvement document and should not be a part of the patient’s medical record.

References


Chylothorax Flowchart

1. **Chylothorax Diagnosis**
   - **High volume output >20 mL/kg/day**
     - NPO with TPN for 7 days
     - CT output <10mL/kg/day at 7 days?
       - YES: Implement Chylothorax diet (continue 4-6 weeks)
       - NO: Initiate Octreotide infusion
         - Remain NPO with TPN
         - CT output <10mL/kg/day at 7 days?
           - YES: Wean Octreotide infusion by 1 mcg/kg/hr q24h
             - Initiate Chylothorax diet
           - NO: Continue Chylothorax diet 4-6 weeks if CT remains <10 mL/kg/day
   - **Low volume output <20 mL/kg/day**
     - Implement Chylothorax diet
     - CT output <10mL/kg/day at 7 days?
       - YES: Continue Chylothorax diet 4-6 weeks
       - NO: Continue NPO with TPN and Octreotide Consider next interventions