MINNESOTA
NEONATAL
PHYSICIANS P.A.

Chylothorax

Diagnostic and monitoring recommendations:

1. Diagnostic thoracentesis (pleural fluid analysis)
    a. WBC>1,000/uL
    b. >70% lymphocytes in cell count
    c. Triglyceride level >110 mg/dL (if being fed)

2. Lab monitoring of documented chylothoraxa
    a. Echocardiogram, MRA, or venogram to evaluate for thrombosis in the left jugular, subclavian, innominate veins, or superior vena cava.
    b. Na, K, Glucose, Ca, BUN, Total protein and albumin at least 2x/week
    c. Antithrombin III level 2 x/week until stable, IF ≥25 ml/kg day or thrombosis
    d. PT, PTT and Fibrinogen weekly
    e. Hct 1x/week
    f. Total IgG level q 1 week
    g. Direct bilirubin q week if NPO > 2 weeks (Nutrition Service recommendation)
    h. Free T4 and TSH q 2 weeks, if using Octreotide

**The above schedule may be altered based on volume of chyle output or clinical stability of the patient.*

Treatment:

1. Thoracostomy drainage to maintain lung inflation and approximate the pleural surfaces.
2. If a thrombosis is present, consider thrombolysis or heparin (Refer to guideline for Thrombosis) and consider Hematology Service consult.
3. NPO and TPN.
4. If thrombus is in the upper venous system, central venous catheters should be placed in the lower body.
5. Fluids should be restricted and diuretics utilized to control edema and reduce chyle flow.
6. Small volume (<25 ml/kg/day) effusions may be replaced with crystalloid or 5% albumin. HOWEVER, if the coagulation status is abnormal, FFP should be used.
7. Large volume (≥25 ml/kg/day) effusions should be replaced with FFP at a ratio of 0.5-0.75 ml FFP/ml of chylous output, as long as clotting function and ATIII levels remain normal.

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8. Additional volume needs may be met with crystalloid solutions or possibly 5% or 25% albumin, IF coagulation status normal.
9. Keep serum albumin concentration ≥2 gm/dL with 25% albumin (5 ml/kg).
10. Keep ATIII concentration >50% in first month, >60% @ 1-3 mo, with ATIII concentrate (75 units/kg IV) or by increasing the ratio of replacement with FFP for chylous output. Consider Hematology consult if there is a thrombus and consideration is being given to heparin therapy.
11. Octreotide therapy should be considered for large volume losses. Start at 1 mcg/kg/hr by continuous infusion and increase by 1 mcg/kg/hour each day to max of 10. Note: It is not compatible with most other things and must be mixed in normal saline or D5W. If separate vascular access is a problem, it may be given subcutaneously or by IV bolus; starting at 10 mcg/kg/day and increasing rapidly to 40 mcg/kg/day divided q 6 hours. Glucose must be monitored closely due to propensity to increase blood glucose. If octreotide works, it usually does so in two weeks. There are significant side effects associated with octreotide and is generally reserved for a last resort therapy.

12. If after 6 weeks* there is no improvement, consider:
    a. Physical or chemical pleurodesis
    b. Thorascopic sealing with fibrin glue
    c. Ligation of the thoracic duct
    d. Pleuroperitoneal shunt (ineffective if RA pressure is >25 mmHg).

**Patients with elevated venous pressure or intravascular clots obstructing the thoracic duct are less likely to respond to conservative management and a surgical solution may be justified earlier.*

13. Documentation of daily output and relevant interventions is highly recommended.
14. Consider Hematology consult, if there is a thrombus and consideration is being given to heparin therapy.

Following resolution of the chylothorax, enteral nutrition is reintroduced with a fat-free formula (ProViMin with polycose). A stepwise protocol is used to increase volumes and then reintroduce short and medium chain fats followed by a final step of reintroduction of long chain fats. Please refer to the ProViMin Feeding Protocol below in Table 1.

ProViMin Recommendations:

The ProViMin regimen is not meant for extended use and should be gradually advanced according to the steps listed below (Table 1 on next page). Advancement of feedings depends on the patient's clinical status and response to the current feeding regimen. The percentage of enteral calories from carbohydrate decreases as the percentage from fat increases, always maintaining a caloric density of 20 cal/oz. The caloric density should not routinely be increased at the first 3 or 4 steps because of the higher osmolality of a formula that is high in carbohydrate and low in fat.

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Advance volume as tolerated until medically ready to introduce and advance enteral fat using MCT (Steps 2 – 5, below). As formula volume is increased, decrease dextrose and protein in TPN. Parenteral lipids can be decreased (0.5-1gm/kg/day minimum) as a source of calories as MCT is increased but should be continued as a source of essential fatty acids until the patient is tolerating step #6 well and is ready to advance to step #7.

Table 1. ProViMin Recommendations

FORMULATION OF ProViMin	COMPOSITION
1. ProViMin + Polycose = 20 cal/oz (no added fat)	**88%CHO, 12%Prot, 0.5%Fat**
2. ProViMin + Polycose + 10% MCT = 20 cal/oz	**77%CHO, 12%Prot, 11%Fat**
3. ProViMin + Polycose + 20% MCT = 20 cal/oz	**67%CHO, 12%Prot, 21%Fat**
4. ProViMin + Polycose + 30% MCT = 20 cal/oz	**57%CHO, 12%Prot, 31%Fat**
5. ProViMin + Polycose + 40% MCT = 20 cal/oz	**46%CHO, 12%Prot, 42%Fat**
6. ProViMin + Polycose + MCT + 4% LCT = 20 cal/oz	**46%CHO, 12%Prot, 42%Fat**
7. ProViMin + Polycose + MCT + 10% LCT = 20 cal/oz	**46%CHO, 12%Prot, 42%Fat**
8. Change to Enfaport	**41% CHO, 14%Prot, 45%Fat**
	(84% of fat is MCT, 16% LCT)

Skimmed Breast Milk:

Low fat breast milk is available from some breast milk banks. The Lactation Consultants can help moms partially skim their own milk to use at the point where adequate LCT tolerance has been demonstrated.

Background

Neonatal chylothorax is a serious, potentially life-threatening problem. Causes include: congenital atresia or hypolasia of the thoracic duct, lymphatic abnormalities lymphangiomatosis), trauma related to birth, placement of central venous catheters, thoracic, cardiac, or neck surgery, especially involving the esophagus, aorta, PDA, or diaphragm, thrombosis of the innominate or subclavian vein, obstruction of the superior vena cava , or increased central venous pressure. If the etiology is post-surgical it typically appears 1-28 days later (avg. 7 days), often coincidental with the introduction of dietary fat.

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The thoracic duct transports up to 150 mL/kg/day of fluid. Flow rate is dependent on diet (any enteral intake at all increases flow, but especially long chain fats). It contains a combination of lymphatic and gut-derived substances. Much dietary fat is absorbed directly into the lymphatic system, however, medium and short chain fatty acids are absorbed directly into the venous system. The thoracic duct is the main vessel for the return of extravasated proteins to the circulation, giving it a major role in fluid homeostasis. The chemical composition of chyle is noted below in Table 2.

Table 2. Content of Chyle

Component	Amount
Total fat	0.4-5 g/dL
Total cholesterol	65-200 mg/dL
Total protein	2.2-5.9 g/dL
Albumin	1.1-4.1 g/dL
Globulins	1.1-3.1 g/dL
Fibrinogen	16-24 g/dL (>25% of plasma)
Anti-thrombin globulin	>25% of plasma
Prothrombin	>25% of plasma
Lymphocytes	400-6000/mm3
RBCs	50-600/mm3
Glucose	Similar to plasma
Electrolytes	Similar to plasma
Calcium	3.4-6 mMol/L
Phosphorus	0.8-4.2 mMol/L
Urea	1.4-3 mMol/L
Pancreatic lipase	0.5-2.4 units/ml
Amylase	50-83 units/ml
AST	22-40 units/ml
Alkaline phosphatase	2-4.8 units/ml
Fat soluble vitamins	??? concentrations