CHYLOTHORAX: PICU PERSPECTIVE

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DISCLAIMER:
INTERPRETATION IS ALL YOURS
The management of chylothorax.
V G Valentine and T A Raffin

*Chest* 1992;102;586-591

[http://chestjournal.chestpubs.org/content/102/2/586.citation](http://chestjournal.chestpubs.org/content/102/2/586.citation)
A Contemporary Review of Chylothorax

Dr A. Talwar, Hans J. Lee. Dept. of Med, Division of Pulmonary and Critical Care Medicine, North Shore University Hospital, NY, USA

There are no evidence-based guidelines to assist in the management of this disease.
Chylothorax: Aetiology, diagnosis and therapeutic options

Emmet E. McGrath, Zoe Blades, Paul B. Anderson
Nutritional Management of Chyle Leaks: An Update
State-of-the-Art Review Article
Chylothorax in Infants and Children
James D. Tutor, MD

Pediatrics 2014; 133:4 722-733
THE BEGINNING: Chyle (Latin for “juice”) Leaks

- **Chylothorax**: Accumulation of fluid in the pleural space, rich in triglycerides and characterized by the presence of chylomicrons.

- **Uncommon**: Most physicians have not cared for patients with a chylothorax.

- **Wide range of Management options**: Diverse etiologies found in the literature can be likened to the numerous anatomic variations of the cisterna chyli and the thoracic duct.
Figure 1. Anatomic pathway of the thoracic duct from its origin, the cisterna chyli, and its major anatomic relationships.
Chylothorax

- Traumatic
  - Non-iatrogenic
    - Knife wound
    - Gun shot wound
    - Childbirth
    - Forceful emesis or cough
    - Blunt trauma to chest wall or vertebrae
  - Iatrogenic
    - Thoracic surgery
    - Head and neck surgery
    - Radiation

- Non-traumatic
  - Malignancy
  - Idiopathic
  - Diseases
    - Sarcoidosis
    - Haemangiomatosis
    - Lymphangioleiomyomatosis
    - Filiariasis
    - Amyloidosis
    - Tuberculosis
    - Retrosternal goitre
    - Superior vena cava obstruction
    - Benign tumour
    - Cardiac failure
    - Transdiaphragmatic movement of chylos ascites
<table>
<thead>
<tr>
<th>Causes</th>
<th>No. of Cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nontraumatic</td>
<td>138 (72)</td>
</tr>
<tr>
<td>Malignant</td>
<td>87 (45)</td>
</tr>
<tr>
<td>Lymphomatous</td>
<td>70 (37)</td>
</tr>
<tr>
<td>Nonlymphomatous</td>
<td>17 (9)</td>
</tr>
<tr>
<td>Nonmalignant</td>
<td>51 (27)</td>
</tr>
<tr>
<td>Idiopathic</td>
<td>26 (14)</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>15 (8)</td>
</tr>
<tr>
<td>Traumatic</td>
<td>53 (28)</td>
</tr>
<tr>
<td>Surgical</td>
<td>48 (25)</td>
</tr>
<tr>
<td>Nonsurgical</td>
<td>5 (3)</td>
</tr>
<tr>
<td>Characteristic or Component</td>
<td>Normal Range</td>
</tr>
<tr>
<td>----------------------------</td>
<td>----------------</td>
</tr>
<tr>
<td>Total Protein</td>
<td>2.2-6.0 gm/dl</td>
</tr>
<tr>
<td>Albumin</td>
<td>1.2-4.2 gm/dl</td>
</tr>
<tr>
<td>Globulin</td>
<td>1.1-3.1 gm/dl</td>
</tr>
<tr>
<td>Total Fat</td>
<td>0.4-6.0 gm/dl</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>65-220 mg/dl</td>
</tr>
<tr>
<td>Triglycerides</td>
<td>&gt;plasma levels</td>
</tr>
<tr>
<td>Glucose</td>
<td>48-200 mg/dl</td>
</tr>
<tr>
<td>Blood urea nitrogen</td>
<td>8-17 mg/dl</td>
</tr>
<tr>
<td>pH</td>
<td>7.4-7.8</td>
</tr>
<tr>
<td>Specific gravity</td>
<td>1.012-1.025</td>
</tr>
<tr>
<td>Electrolytes</td>
<td></td>
</tr>
<tr>
<td>Sodium</td>
<td>104-108 mEq/L</td>
</tr>
<tr>
<td>Potassium</td>
<td>3.8-5.0 mEq/L</td>
</tr>
<tr>
<td>Chloride</td>
<td>85-130 mEq/L</td>
</tr>
<tr>
<td>Calcium</td>
<td>3.4-6.0 mEq/L</td>
</tr>
<tr>
<td>Enzymes</td>
<td></td>
</tr>
<tr>
<td>AST</td>
<td>22-40 U/ml</td>
</tr>
<tr>
<td>ALT</td>
<td>5-21 U/ml</td>
</tr>
<tr>
<td>Amylase</td>
<td>50-83 U/ml</td>
</tr>
<tr>
<td>Cell count</td>
<td></td>
</tr>
<tr>
<td>Lymphocytes</td>
<td>400-6,800/µl</td>
</tr>
<tr>
<td>Erythrocytes</td>
<td>50-600/µl</td>
</tr>
</tbody>
</table>

*AST = aspartate aminotransferase; ALT = alanine aminotransferase.
## Components of Chyle

<table>
<thead>
<tr>
<th>Component</th>
<th>Concentration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calories</td>
<td>200 kcal/L</td>
</tr>
<tr>
<td>Lipids</td>
<td>5–30 g/L</td>
</tr>
<tr>
<td>Protein</td>
<td>20–30 g/L</td>
</tr>
<tr>
<td>Lymphocytes</td>
<td>400–6800/mm</td>
</tr>
<tr>
<td>Erythrocytes</td>
<td>50–600/mm</td>
</tr>
<tr>
<td>Sodium</td>
<td>104–108 mMol/L</td>
</tr>
<tr>
<td>Potassium</td>
<td>3.8–5.0 mMol/L</td>
</tr>
<tr>
<td>Chloride</td>
<td>85–130 mMol/L</td>
</tr>
<tr>
<td>Calcium</td>
<td>3.4–6.0 mMol/L</td>
</tr>
<tr>
<td>Phosphate</td>
<td>0.8–4.2 mMol/L</td>
</tr>
</tbody>
</table>
Diagnostic Features of Chylous Effusions

- **Appearance**
  - Commonly milky, serous/sanguineous
- **Odor**
  - Odorless
- **Supernatant**
  - Opalescent
- **Cell count**
  - Lymphocytes mostly
- **Culture**
  - Sterile
- **Triglycerides**
  - > 110 mg/dl
- **Lipoprotein**
  - Chylomicrons
- **Ingestion of cream**
  - Opacifies clear effusion
### Table 1: Pleural fluid criteria for the diagnosis of chylothorax and pseudochoylothorax.

<table>
<thead>
<tr>
<th></th>
<th>Triglyceride</th>
<th>Cholesterol</th>
<th>Chylomicron</th>
<th>Cholesterol crystals</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Chylothorax</strong></td>
<td>&gt; 1.24 mmol/l</td>
<td>&lt;5.18 mmol/l</td>
<td>Present</td>
<td>Not seen</td>
</tr>
<tr>
<td></td>
<td>(110 mg/dl)</td>
<td>(200 mg/dl)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Pseudo-choylothorax</strong></td>
<td>&lt; 0.56 mmol/l</td>
<td>&gt;5.18 mmol/l</td>
<td>Absent</td>
<td>Often seen</td>
</tr>
<tr>
<td></td>
<td>(50 mg/dl)</td>
<td>(200 mg/dl)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
MANAGING CHYLOTHORAX

• Once the physician diagnoses chylothorax, he/she must carefully evaluate the setting in which it developed.

• Surgical causes are obvious.

• However, the nonsurgical/traumatic causes require special attention.

• Consequently, the first step in the management of chylothorax demands a review of the history and physical examination.
THE PROBLEM

- No RCTs
- Infrequent occurrences
- Diverse etiologies
- Varied presentations
Initial Approach

• Decompression of the pleural space and the thoracic lymphatics as chylous effusion may compress intrathoracic structures and embarrass respiration.

• Single or Multiple thoracenteses/or continuous tube drainage.

• Tube drainage is considered more effective
  (A) Pleural surface apposition to the fistula may accelerate healing.
  (B) Provides a means of accurately monitoring the rate of chyle leakage.
REAL CONCERNS

• Up to 3 L of chyle may drain daily.

• Large amounts of fluid, electrolytes, protein, fat, fat-soluble vitamins, and lymphocytes, predominantly the T-cell variety may be lost resulting in severe nutritional depletion and immunodeficiency.

• For replacement, monitoring the patient's weight, serum albumin, total protein, absolute lymphocyte count, and electrolytes is required.
NUTRITIONAL MANAGEMENT OF CHYLE LEAKS: The Goals

• 1. Decrease production and flow of chyle: Provide symptom relief, avoid aggravating the leak, and allow closure of the leak if possible.

• 2. Replenish fluid and electrolytes losses.

• 3. Prevent malnutrition: aid in maintaining or repleting nutritional status.
Options for nutritional management

- A low fat or fat free oral diet
- Enteral nutrition with specialized Formula
- TPN without oral intake
- Some combination of these
WHICH ONE IS THE BEST?

• All recommendations based on isolated cases and cohorts of patients, and the effect of different oral foods, enteral formulas or fluids on chyle flow.

• No consensus as to the best type of regimen, how long nutrition management should be pursued, or what constitutes an acceptable amount of chyle output.
ORAL DIET

- Well nourished and able to take food by mouth: Fat-free oral diet
- Fat is a great calorie source
- Fat severely limited > Additional calories obtained from larger volumes of other food (more meals and snacks) throughout the day.
- Virtually impossible to remove all fat from the diet. Many fruits, vegetables and even “fat free” products contain traces of fat (“fat free” = <0.5 g/serving).
ORAL DIET

• Nutritional status monitored carefully in patients on a fat free diet.
• Fat-soluble vitamins and/or EFA may need to be supplemented.
• MV, mineral supplement may also be necessary.
• Some may need supplemental nutrition support, such as nocturnal infusion of a low or fat free enteral formula.
MCT BASED DIET

- MCT are frequently ordered for the treatment of chyle leaks—in fact, what the dietitian typically receives is an order for a "MCT diet." (A MYTH?)
- Do not require transport via the lymph system.
- Hydrolization to MCFA occurs rapidly, allowing absorption across the brush border where the MCFA then bind with albumin and are transported directly to the liver via the portal vein.
ANY CONCERN WITH MCT?

• Some MCT up to 20% may find their way into the lymphatic system and make up part of the lymph fluid, especially in the setting of high MCT intake, or high total fat intake.
MCT COMPOSITION

- A mixture of caproic (C6:0 @ 1 to 2%), caprylic (C8:0 @ 65 to 75%), capric (C10:0 @ 25 to 35%), and lauric acids (C12:0 (1 to 2%).
- MCFA for MCT obtained by hydrolysis of coconut oil, then fractionated, and reesterified with glycerol into MCT.
- Coconut oil and MCT are not the same, as only 66% of coconut oil is MCT—the rest is long chain fat (i.e. coconut oil should not be used in the treatment of chyle leaks).
Dietary Modification based on MCT

• Can address nutritional needs.
• MCTs as a source of fat is an invaluable adjunct.
• Variable success rates.
• MCTs have a variety of substrates: Trioctanoin may be the preferable MCT substrate for the modified diet.
• Thoracic duct flow is minimized to promote healing of the leak.
Incorporating MCT into the Diet: A CHALLENGE

• Sip or eat slowly at least initially
• Start with small volume. Taken as small “shots”.
• Add to a fat free beverage of choice.
• Flavorings can be added.
• Consider diluting with equal volume of water or other fat free beverage, fat free hot cereals, mashed potatoes, vegetables, pasta, soups, salad dressings, applesauce or other fruit sauces.
Some authors believe that the optimal approach at the outset includes a regimen of:

(A) No oral intake
(B) TPN
(C) Tube thoracostomy drainage
## Fat-Free Oral Supplements Options: Nutrients per Serving

<table>
<thead>
<tr>
<th>Product</th>
<th>Calories</th>
<th>Protein</th>
<th>Fat</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enlive® (Abbott®)</td>
<td>200</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Resource® Breeze (Nestle®)</td>
<td>250</td>
<td>9</td>
<td>0</td>
</tr>
<tr>
<td>NUTRA/Shake® Fruit Plus (NUTRA/Balance®)</td>
<td>200</td>
<td>6</td>
<td>0</td>
</tr>
</tbody>
</table>
### Examples of Fat Free Protein Sources

<table>
<thead>
<tr>
<th>Product</th>
<th>Serving Size</th>
<th>Protein (g)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Egg Beaters®</td>
<td>¼ c</td>
<td>6</td>
</tr>
<tr>
<td>Better n’Eggs®</td>
<td>¼ c</td>
<td>5</td>
</tr>
<tr>
<td>Egg whites</td>
<td>2 tsf</td>
<td>7</td>
</tr>
<tr>
<td>Fat free luncheon meat</td>
<td>1 oz</td>
<td>6</td>
</tr>
<tr>
<td>Fat free milk/milk powder</td>
<td>8 oz</td>
<td>8</td>
</tr>
<tr>
<td>Non-fat cheese</td>
<td>1 oz</td>
<td>8</td>
</tr>
<tr>
<td>Non-fat yogurt (plain)</td>
<td>8 oz</td>
<td>12</td>
</tr>
<tr>
<td>High protein gelatin</td>
<td>½ cup</td>
<td>12</td>
</tr>
</tbody>
</table>
SURGICAL INTERVENTION

- In the paediatric population conservative management steadily reduces the chyle loss which may continue for weeks or months, provided the nutritional state is maintained.
- Imminent nutritional deterioration or the re-accumulation of chyle following the reinstigation of dietary fat is an indication for Surgical management.
- Selle and associates
  (A) Average daily loss >1,500 ml in adults or 100 ml per year of age in children for a 5 day period
  (B) When the chyle flow has not diminished by 2 weeks of conservative management
SURGICAL OPTIONS

• Diagnostic or therapeutic Thoracotomy to locate the site of disruption and to ligate/oversew the injury.
• Thoracic duct ligation.
• Pleuroperitoneal shunting.
• In situations where it may be impossible to identify the duct (malignancy or radiation fibrosis obscuring the duct) other modalities, including pleurodesis and pleurectomy, may be used.
Recommended treatment pathway for chylothorax.

1. Pleural effusion
   - Unilateral or bilateral
   - Thoracentesis
   - Clinical suspicion for chylothorax
     - (White or yellow aspirate, lymphoma, trauma, post thoracic surgery)

2. ↑ Triglycerides, ↓ Cholesterol, + Chylomicrons
3. Chylothorax → Transudate
4. Investigate and treat concomitant illness eg. cardiac or liver failure
5. Investigate and treat aetiology eg. chemoradiotherapy for malignancy

Conservative treatment

- Nil by mouth
- Oral medium chain triglycerides
- Total parenteral nutrition
- Intercostal drain insertion
- Talc pleurodesis
- Somatostatin

If drains > 1.5L/24 hr or > 1L/day x 5 day or Persistent leak for > 2 weeks or Nutritional or metabolic compromise

Surgical treatment

- Chylothorax post oesophagectomy
- Ligation of leak under direct visualisation (VATS/open surgery)
- Mass ligation of thoracic duct
- Surgical pleurodesis
- Pleurectomy
- Pleuroperitoneal shunt
CONCLUSIONS

• Because of its diverse etiology and infrequent occurrence, the approach to chylothorax remains unsettled in the literature.

• Proper management will depend on the initial nutritional state of the patient, the cause and severity of the chylothorax, and, in rare instances, the anatomy of the thoracic duct.

• Familiarity with the therapeutic options, along with appropriate timing for surgical intervention, will be required to prevent the complications of malnutrition and infection from persistent chyle loss when other modes of therapy are failing.
Globalize the Evidence
Localize the Decision Making
Thank you for your attention

- 'Half of what you are taught as medical students will in ten years have been shown to be wrong, and the trouble is, none of your teachers knows which half…’

- S Burwell, Dean of Harvard Medical School from 1935 to 1949
THANK YOU FOR A PATIENT HEARING