Continuing Education Series

Management of Chylothorax in Children

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Chylothorax is a collection of a white milky drainage from the lymphatic system within the thoracic cavity that can impair respiratory status as it accumulates. Management of this uncommon complication of thoracic surgery can include: total parenteral nutrition (TPN), low-fat enteral nutrition, thoracentesis to remove the chylous fluid, and surgical ligation of the thoracic duct, as well as octreatide infusion to control splanchnic blood flow. Nursing management of these patients can be complex and challenging.

Chylothorax is an uncommon complication of thoracic surgery associated with significant morbidity and extended hospital stay. Incidence of this complication ranges from 0.6% to 2.5% of children undergoing cardiac surgery. The management of these patients includes drainage of the chylous effusion and dietary limitation of fat intake, as well as other surgical and medical therapy. The difficulties in the management of these patients make treatment of the child with a chylothorax a nursing challenge.

Chylothorax is a collection of lymphatic fluid in the pleural or mediastinal space. It most commonly occurs as a postoperative complication of cardiothoracic surgery but can also appear after thoracic surgery of the esophagus or chest blood vessels (see Table 1). Chylothorax is especially likely to occur in children with elevated pressure in the systemic venous system of the upper body, such as those children who have undergone hemifontan, bi-directional Glenn or Fontan cardiac procedures. The elevated pressure creates a relative obstruction of flow, which prevents the drainage of chyle from the thoracic duct into the left subclavian vein. Vascular thrombus in the superior vena cava or subclavian vessels can also create an elevated systemic venous pressure leading to chylothorax or can directly obstruct chylous flow from the thoracic duct to the subclavian. In addition, there are case reports of congenital and spontaneous chylothorax.

Chyle is an opaque milky white fluid consisting of protein, fat, lymphocytes, and electrolytes absorbed through the gut into the lymph channels of the gastrointestinal tract. The lymph channels drain into the cisterna chyle and then travel up the thoracic duct to re-enter the vascular system near the junction of the left subclavian and left internal jugular veins (see Figure 1). The position of the thoracic duct near the mediastinal structures — heart, aorta, esophagus, and systemic veins — makes it clear how iatrogenic trauma during thoracic surgery could occur.

The composition of chyle is key in determining appropriate patient management of a persistent effusion (see Table 2). Studies in both adults and children show chyle is predominantly protein with a high cell count of greater than 1,000 cells/l. The cell count consists of predominantly lymphocytes, frequently greater than 90%. The fluid is sterile and somewhat bacteriostatic due to the presence of so many white blood cells. If the child has received enteral formula or food, the fluid will also contain elevated triglycerides at greater than 110 mg/dl. These features — cell count, percentage of lymphocytes, and triglyceride level — are helpful in differentiating serous effusion and empyema fluid from chylous drainage. The composition of chyle also points to the management of a chylous effusion.

Because chyle fluid originates in the gut and chyle flow increases dramatically after enteral intake, management of patients with chylothorax has focused on altering dietary intake. If this is not successful, other approaches include surgical management via thoracic duct ligation, pleurodesis, or pleuroperitoneal shunt. More recently, octreatide infusion has also been used.

Dietary Management

The goal of nutrition therapy in chylothorax is to reduce the flow of chyle through the thoracic duct. Since it is mainly long chain fatty acids that are absorbed from the intestines via lacteals and enter the central circulation at the thoracic duct (Shils, Olson, & Shike, 1994), enteral intake of long chain fats must be severely limited. Thus, dietary management includes complete gut rest with parenteral nutrition, relatively fat-free enteral feeding, or very low long-chain triglyceride-high medium chain triglyceride (MCT) enteral feeding (Beghetti et al., 2000; Buttiker, Fanconi, & Burger, 1999). Medium chain fatty acids (6-12 carbons in length) are absorbed directly into the portal system and do not enter the lymphatic system (Shils et al., 1994).

The patient must be provided with adequate calories, fluid, electrolytes, and protein regardless of feeding method. It is also important to provide enough essential fatty acids (the long chain fatty acids linoleic acid and linolenic acid) to prevent essential fatty acid (EFA) deficiency. The American Academy of Pediatrics (AAP) recommends that at least 3% of daily calories come from essential fatty acids (AAP, Committee on Nutrition, 1998). Others report adequate EFA if linoleic acid supplies 1%-

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The CE Posttest can be found on pages 296-297.

Table 1. Causes of Chylothorax

1. Iatrogenic trauma to the thoracic duct during surgery
   - Cardiac
   - Esophageal
   - Diaphragm
   - Vascular procedures in the upper body
2. Elevated systemic venous pressure from
   - Vessel thrombosis (SVC or subclavian)
   - Single ventricle physiology
3. Congenital
4. Idiopathic or spontaneous

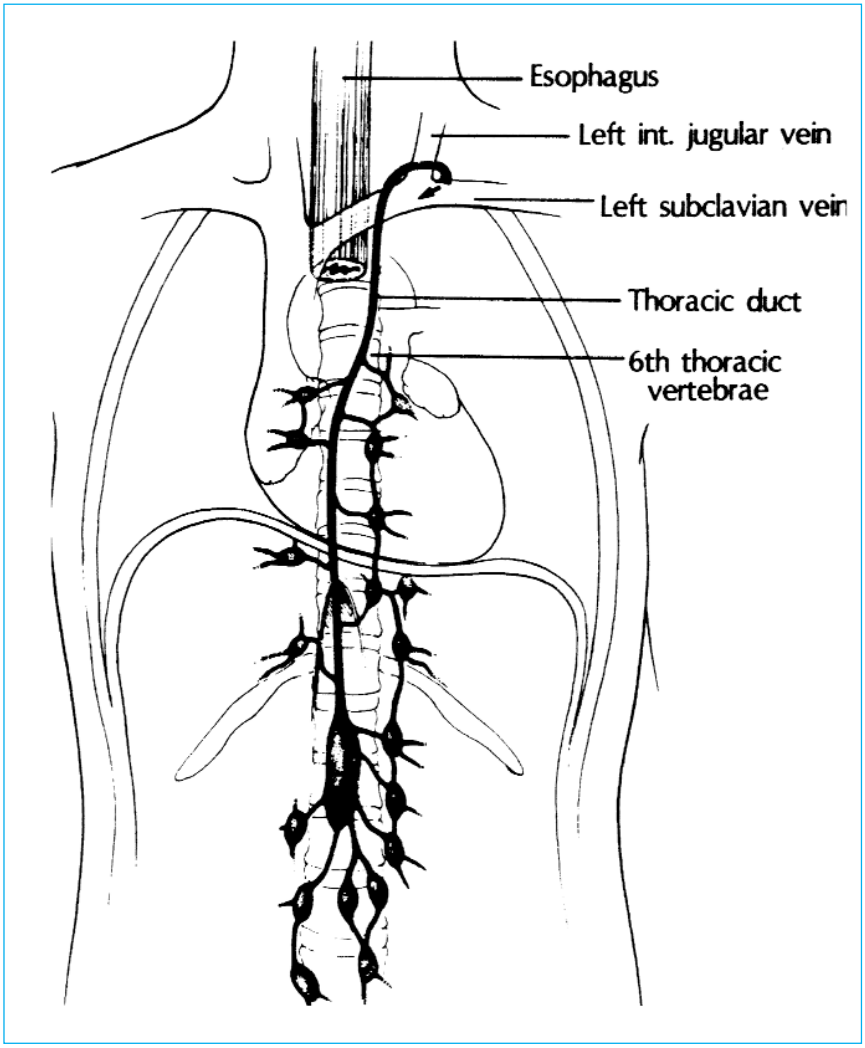
From: Buttiker, 1999

Table 2. Characteristics of Chyle

- Lymphocytes > 80-90%
- Proteins – Albumin
  Immunoglobulins
  Fibrinogen
- Cells > 1,000 / ml
- Triglycerides > 110/ dl (higher than plasma)
- PH 7.4-7.8
- Electrolytes- similar to plasma
- Sterile

Note: From Buttiker et al., 1999; Teba et al., 1985.

Figure 1. Lymphatic System



Note: Used with permission from Teba, L., Dedhia, H.V., Bowen, R., & Alexander, J.C. (1985). Chylothorax review, Critical Care Medicine, 13(1), 50.

2% of total calories and linolenic acid supplies 0.54% of total calories (Samour, Helm, & Lang, 1999). Soybean, safflower, and corn oils are high in linoleic acid; soybean and canola oils are high in linolenic acid.

When total parenteral nutrition (TPN) is the feeding method, adequate essential fatty acids are easily provided (Baker, 1997). The source of fat in parenteral preparations available in the United States and elsewhere is either soybean oil or a mixture of soybean and safflower oils (Baker, 1997). Since the fat in TPN is delivered directly into the blood stream, it never enters the lymphatic system and, therefore, has no effect on the thoracic duct.

Although TPN allows complete gut rest and may be necessary, especially in the unstable patient, it is often preferable to try low fat or non-fat enteral feeding first. Nguyen and colleagues (1995) report similar durations of lymph leakage in both TPN and low-fat enteral feedings. If a formula is required because the patient is an infant and/or is being tube-fed, there are several choices available. Portagen® (Mead Johnson) has been used frequently because 85% of the fat in Portagen is from MCT. It delivers 3.5 grams of protein per 100 calories and 6% of calories from long chain fat in the form of corn oil (Mead Johnson, 2001). However, a recent report of an infant death associated with *Enterobacter sakazakii* (Centers for Disease Control & Prevention [CDC], 2002) traced to Portagen powder has generated new guidelines from the Federal Drug Administration, the CDC, and the American Dietetic Association on the use of powdered formula in health care facilities. It is now recommended that non-sterile powders be avoided when possible.

Lipisorb®, a sterile liquid formula also made by Mead Johnson, has 86% of fat in the form of MCT. Six percent

Table 3. Formula Comparison per 100 calories

	Portagen	Lipisorb	Tolerex	Vivonex	Nutren 2.0	Infant formula guidelines
Kilocalories	100	100	100	100	100	100
Protein g	3.5	4.2	2.1	3.8	4.0	1.7-3.4
Carbohydrate g	11.5	11.8	23.0	21.0	9.8	9-13
Fat g	4.8	4.18	0.15	0.28	5.03	4.4-6.4
% MCT	86	85	0	0	75	
LCT (gms)	0.672	0.627	0.15	0.28	1.2	
Linoleic acid (mg)	370	320				350
Linolenic acid (mg)	8	50				77
Vit. A IU	780	468	278	250	400	200-500
Vit. D IU	78	37.5	22	20	28	40-100
Vit. E mg alpha-TE	2.08	1.88	1.14	1.01	1.39	0.5-5.0
Vit. K mcg	15.6	7.5	4.4	4.0	5.0	1-25
Vit. C mg	8.1	5.6	3.3	6.0	14	6-15
Vit. B1 mcg	156	140	83	150	200	30-200
Vit. B2 mcg	188	160	94	170	240	80-300
Vit. B3 mcg	2100	1870	1100	2000	2800	550-2000
Vit. B6 mcg	210	187	110	200	400	30-130
Folic acid mcg	15.6	37.5	22	40	54	11-40
Pantothenic acid mcg	1050	930	560	1000	1400	300-1200
Vit. B12 mcg	0.63	0.56	0.33	0.6	0.8	0.08-0.7
Biotin mcg	7.8	28	17	30	40	1-15
Choline mg	13.3	15.6	4.1	20	45	7-30
Calcium mg	94	62.5	56	50	67	50-140
Phosphorus mg	70	62.5	56	50	67	20-70
Magnesium mg	20	25	22	20	26.8	4-17 mg
Zinc mg	0.94	1.25	0.83	1.1	1.4	0.4-1.0
Iron mg	1.88	1.125	1.0	0.9	1.2	0.2-1.65
Copper mcg	156	125	110	100	140	60-160
Manganese mcg	125	187.5	110	100	260	1-100
Iodine mcg	7	9.37	8.9	8.0	10	8-35
Sodium mg	55	100	47	60	65	25-50
Potassium mg	125	125	117	95	96	60-160
Chloride mg	86	162.5	95	85	93.8	50-160
Selenium mcg	0	4.38	3.9	3.5	4.0	1.5-5.0

Note: From Mead Johnson, 2001; Nestle Clinical Nutrition, 2002; Novartis Nutrition, 1997; Raiten et al., 1998.

of calories come from long chain fat in the form of soybean oil. It provides 4.2 grams of protein per 100 calories (Mead Johnson, 2001). This amount is twice the protein found in regular infant formula, but extra protein is needed if chest tubes are present. Lipisorb has a relatively high sodium content (100 mg or 4.3 mEq per 100 calories) (Mead Johnson, 2001). However, many patients with chylothorax have chest tubes and are on diuretics, both of which mean increased sodium losses. Lipisorb provides 40 calories per ounce, but it may be diluted with sterile water to appropriate calorie levels. (Trained personnel should dilute formula under aseptic conditions.)

Other very low fat formula powders that may be considered are Tolerex ® (Novartis) and Vivonex TEN® (Novartis). Tolerex provides only 1% of calories from fat and Vivonex only 3 percent (Novartis Nutrition, 1997). With these formulas most patients need additional fat and protein. Fat or protein modules can be added to formula, and/or intravenous lipid can be administered to insure adequate EFA. These additions not only make a complicated nutrition plan, but also increase the risks of infection and formula contamination.

Some formulas that are lower in MCT fat than those discussed above but higher than standard formulas are Nutren 2.0® (75% MCT), Pregestimil® (55% MCT), or Alimentum® (50% MCT) (Nestle Clinical Nutrition, 2002; Mead Johnson, 2001). These formulas are more appropriate as a transition back to standard formula than as therapy for chylothorax. They may provide too much LCT and not allow the leak to heal. An evaluation by a Registered Dietitian in selecting an appropriate enteral formula is advisable (see Table 3).

For those patients who are eating solid foods by mouth, providing a balanced, very low fat diet with adequate calories has become more manageable. Lipisorb is vanilla-flavored and may be accepted by some patients as an oral supplement. When unflavored formula is used (Portagen, Vivonex

Table 4. Clear Liquid, Non-Fat Supplements

Supplement	Manufacturer	Packaged as	Flavors	Calories	Protein
Boost Breeze®	Mead Johnson	8 oz can	Tropical Fruit Mixed Berry	160	8
Enlive®	Ross Laboratories	8.1 oz brick pack	Apple Peach	300	10
NuBasics®	Nestle	5.5 oz can	Tangy Orange Sweet Berry	163	6.5
Resource Fruit® Beverage	Novartis	8 oz brick pack	Orange Peach Wild Berry	250	9
Resource® Nutritious Juice Drink	Novartis	6 oz brick pack	Orange Apple Cranberry Golden Tropical	210	6

Note: From: Mead Johnson, 2001; Nestle Clinical Nutrition 2002; Novartis Nutrition, 1997; Ross Nutrition, 2003.

TEN, Tolerex), adding a drop of non-alcoholic food flavoring makes the formula more palatable. Some formula manufacturers also make flavoring packets.

Manufacturers of medical nutritional products also make juice-type supplements. These products contain no fat but do provide vitamins, minerals, and protein. Most of them are not available in retail markets, but may be ordered from the manufacturer. They are typically less expensive than traditional supplements like Pediasure®, Kindercal®, Nutren Jr.®, etc. Boost Breeze® by Mead Johnson is an exception and is in many supermarkets and pharmacies. See Table 4 for other choices.

Many non-fat dairy products that provide a good source of protein (non-fat milk, non-fat yogurt) are available. Another non-fat source of protein is egg white. Egg whites can be obtained from fresh whole eggs (always well cooked) or purchased as a dry powder or a pasteurized liquid. The powder can be added to other foods to increase protein as needed. The liquid pasteurized egg white can be used as a substitute for whole egg in cooked dishes.

With the exception of coconut and avocado, fruits and vegetables are very low in fat. A healthy diet includes 5 servings of fruits and vegetables a day. There are many low fat and non-fat breads and cereals as well as baked products now available. Because meat, fish, and poultry contain fat that is impossible to remove, it is important that each individual be evaluated for an acceptable level of dietary fat before these animal products are allowed. It is helpful to provide families with a guideline of an acceptable amount of fat in grams per day. Giving the family information on selecting lower fat meat, fish, and poultry, as well as low fat preparation techniques, can help the family adhere to the fat restriction. The family may also benefit from instruction on reading food labels and determining fat content. See Table 5 for information on foods.

The amount of fat allowed should meet minimum essential fatty acid needs. Typically, patients are on the restricted fat diet or high MCT formula for 4-8 weeks (Nguyen, Shum-Tim, Dobell, & Tchervenkov, 1995). They should be followed carefully for signs of essential fatty acid deficiency, especially if the low-fat diet is needed beyond 4-6 weeks. EFA deficiency has been found in infants and children receiving a fat-free diet for greater than 3 weeks (Baker, 1997). Symptoms of a deficiency are scaly skin, delayed wound healing, poor growth, diarrhea, platelet dysfunction, and hair loss (Baker, 1997). An increased ratio of serum trienes to tetraenes (> 0.4) indicates EFA deficiency (AAP, Committee on Nutrition, 1998).

Medical Management

In addition to controlling effusion volume via dietary management, effective drainage of the effusion via thoracentesis or more commonly chest tube placement is necessary to optimize respiratory status. One of the major concerns then becomes the loss of key proteins and fluid through the drainage fluids. Regular laboratory assessment of albumin and immunoglobulin levels will indicate whether supplementation of these proteins is needed in an individual case. Some centers replace cc for cc losses while there is significant volume output. Immunoglobulin replacement with IVIG supports patients through a period when they are at risk for superimposed infection from loss of white blood cells, antibodies, and proteins needed for adequate recovery from surgery.

Surgical Management

Surgical management of a chylothorax remains controversial and quite variable from hospital to hospital. Some centers will ligate the thoracic duct after a certain number of weeks of drainage, while others use volume of drainage and rate of decline in drainage as determining factors to plan surgery. Some hospitals use pleurodesis, the stripping of the pleura off the surface of the lung, to control fluid drainage. Still others use irritant agents (such as talc) to sclerose the pleura to the lung. Also reported in the literature is placement of a pleuroperitoneal shunt to shift the fluid from the thoracic cavity to the abdominal cavity where it can be reabsorbed. (Wolff, 1999) This approach prevents the problem of electrolyte and protein loss. This technique could be successful or at least minimally helpful depending on the volume of fluid shunted but could create an ascites like picture in the patient.

New Techniques

The newest medical therapy reported in small groups of patients is the use of octreotide as a continuous

Table 5. Food Guidelines

Food Group	Recommended Foods	Foods to Avoid	Tips
Breads, cereals, rice, and pasta	White breads, cereals, rice, barley, and pastas Fat-free crackers and cookies	Bread with eggs or cheese, granola-type cereal, cereals with nuts, biscuits, waffles, pancakes, croissants, muffins, doughnuts, high fat crackers	Try French bread, pita bread, plain bagels, bread sticks. Puffed Rice and Rice Krispies® are very low fat.
Fruits	All fresh, frozen, canned, or dried fruit; fruit juices	Avocado, coconut	Use fresh or dried fruit as snacks.
Vegetable	Fresh, frozen, or canned vegetables	Vegetables with added fat, cream or cheese sauce, fried vegetables	Cook vegetables in broth or sprinkle with herbs and spices to add flavor.
Milk, yogurt, and cheese	Skim milk, non-fat cheese, nonfat yogurt or cottage cheese, fat-free sour cream or cream cheese	1%, 2% and whole milk; buttermilk, chocolate milk, cream, regular, hard, and processed cheeses	Substitute skim milk, evaporated skim milk, or nonfat yogurt for whole milk and cream. Stoneyfield Farm® makes nonfat yogurt with no aspartame.
Snacks, sweets, condiments, and beverages	Fat-free broth or soups, fruit ice, popsicles, gelatin, angel food cake, graham crackers, and nonfat desserts; honey, jams, jellies, syrups, and hard candy; soda, fruit drinks, other nondairy drinks	Cream or cheese sauces or soups, gravy, mayonnaise; cakes, cookies, pies, and ice cream; coconut, chocolate, creamed candy, candy with nuts; chips, buttered popcorn	Try lemon juice, vinegar, garlic or onion powder, fat-free margarine, dressings, mayonnaise, marshmallows.
Meats, poultry, fish, beans & peas, eggs, and nuts	Lean beef sirloin, round, chuck or veal; lean pork, tenderloin or chops; lean lamb chops or leg; poultry without skin; fresh or frozen fish, canned fish in water; egg whites, tofu, or beans made without added fat	Fried, fatty, or heavily marbled meat, poultry, or fish (ground beef, pork, lamb, ribs, corned beef, sausage, chicken thigh); canned fish packed in oil; luncheon meat (bologna, salami, hot dogs); pizza, nuts, peanut butter	Broil, roast, grill, or boil meats, poultry, and fish. Trim all visible fat before cooking. Use natural juice instead of with sauces and gravies. BallPark Brand makes a fat-free hot dog.

Note: From Pennington, 1994.

or intermittent infusion. Octreotide is known to decrease splanchnic, hepatic, and portal blood flow, thereby decreasing the volume of lymph fluid produced and, ultimately, thoracic duct flow. Octreotide inhibits the absorption of triglycerides and decreases acetylcholine release in the gut. Acetylcholine is known to increase lymph flow so reduced acteylcholine would result is decreased lymph flow. The case reports suggest 1-4 mcg/kg/hr infusion when used in conjunction with dietary techniques appeared helpful in reducing lymph volume (Cheung, Leung, & Meimei, 2001; Pettitt, Caspi, & Borne, 2002; Pratap, Slavik, Ofoe, Onuzo, & Franklin, 2001). Anecdotal reports of side effects include vomiting and other gastrointestinal issues related to decreased blood flow. The pharmacology literature also reports risk of arrhythmias including bradycardia, conduction abnormalities such as heart blocks, and prolongation of the QT interval. Because many patients with chylothorax are cardiac, they should be carefully monitored for these possible side effects.

Conclusion

Management of the uncommon complication of chylothorax in a child challenges nurses to not only care for their underlying disease but also to monitor the effectiveness of TPN or low-fat enteral nutrition in reducing the chylous effusion. Nurses must also conduct routine respiratory assessment and chest tube management. In addition, replacement of protein or immunoglobulin losses may be required. Consultation with a pediatric dietician who can recommend the best formula preparations and possible food choices is key. Identifying flavorings for less palatable formulas or seeking low-fat alternative foods is important to ensure good nutrition for the more picky eaters.

Managing the child with chylothorax presents still other difficulties. The lack of consensus regarding when to move beyond thoracentesis or chest tube placement to remove the fluid and ini-

tiate surgical ligation of the thoracic duct or other surgical techniques complicate the nurses' role in educating patient/family about the plan of care.

Moreover, the recent introduction of octreotide infusion requires nurses to gain familiarity with a new approach. Nonetheless, octreotide infustion offers hope that new techniques will reduce the length of hospitalization and the complications that can occur as a result of protein, immunoglobulin and fluid losses and may ease the challenge of nursing management of these complex pediatric patients.

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