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Nutritional management of postoperative chylothorax in children with CHD

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Abstract

Introduction: Chylothorax after congenital cardiac surgery is associated with increased risk of malnutrition. Nutritional management following chylothorax diagnosis varies across sites and patient populations, and a standardised approach has not been disseminated. The aim of this review article is to provide contemporary recommendations related to nutritional management of chylothorax to minimise risk of malnutrition. *Methods:* The management guidelines were developed by consensus across four dietitians, one nurse practitioner, and two physicians with a cumulative 52 years of experience caring for children with CHD. A PubMed database search for relevant literature included the terms chylothorax, paediatric, postoperative, CHD, chylothorax management, growth failure, and malnutrition. *Results:* Fat-modified diets and nil per os therapies for all paediatric patients (<18 years of age) following cardiac surgery are highlighted in this review. Specific emphasis on strategies for treatment, duration of therapies, optimisation of nutrition including nutrition-focused lab monitoring, and supplementation strategies are provided. *Conclusions:* Our deliverable is a clinically useful guide for the nutritional management of chylothorax following paediatric cardiac surgery.

Postoperative chylothorax occurs in about 3.8% of paediatric patients following cardiac surgery and is associated with increased morbidity and mortality.[1] Chyle is composed of predominantly fat, protein, lymphocytes, and electrolytes. This typically accounts for 200 kcal/L in a healthy patient. The clinical consequences of continued chylous drainage include electrolyte disturbances, risk of infection, and nutrient losses compounding the existing increased energy demand secondary to surgery, critical illness, and postoperative recovery. Chylothorax amplifies the underlying burden of growth failure inherent to many cardiac lesions.[1,2]

Materials and methods

The Chylothorax Work Group was formed in October 2020. Members represent 22 centres and consist of more than 60 multi-disciplinary providers: physicians, surgeons, advanced practice providers, and dietitians.

Using the Chylothorax Work Group infrastructure, a group of experts were identified to develop contemporary and clinically relevant guidance related to nutritional management of paediatric postoperative chylothorax. The experts included four dietitians (KF, MH, AT, and PV), one Nurse Practitioner (MW), and two cardiac intensive care physicians (EG and DB). The dietitian-specific expertise accumulates to 38 years of experience caring for children with CHD.

Literature was reviewed using the PubMed search terms chylothorax, paediatric, postoperative, CHD, chylothorax management, growth failure, and malnutrition between the years 2001 and 2021.

Results

In the management of chylothorax, the overarching nutritional goals are to decrease chylous drainage, maintain adequate volume and electrolyte status, and prevent further malnutrition. The thoracic duct and its lymphatic tributaries transport about 4 L of chyle per day.[3] Chyle is composed of lipids, proteins, fat-soluble vitamins, lymphocytes, and electrolytes (Table 1). The continuous loss of lymphatic fluid may result in the loss of nutrients, proteins, immunoglobulins, coagulation factors, and vitamins leading to respiratory compromise, nutritional deficiency, infections, haematologic complications, and metabolic derangements[4] in an already

Table 1. Composition of chyle

Relative density	1.012–1.015
pH	7.4–7.8
Color	Milky (colorless if NPO)
Sterile	Yes
Bacteriostatic	Yes
Fat (g/L)	5–30
Protein (g/l)	20–30
Albumin	12–42
Globulin	11–31
Albumin:globulin ratio	3:1
Fibrinogen (mg/L)	160–240
Glucose (mmol/L)	2.7–11.1
Cell count (per dl)	
Lymphocytes	40,000–680,000
Erythrocytes	5,000–60,000
Electrolyte concentration (mmol/L)	
Sodium	104–108
Potassium	3.8–5.0
Chloride	85–130
Calcium	3.4–6.0
Phosphate	0.8–4.2

vulnerable population. The prevalence of chylothorax is increased in the most nutritionally at-risk populations such as neonates, infants, single-ventricle anatomy/physiology, with or without arch reconstruction and those with genetic syndromes.[1,5] With the significant risk for malnutrition in this population, a standardised approach to management in conjunction with close monitoring from a registered dietitian in collaboration with the multidisciplinary team is paramount.

Fat-modified diet therapies

In growing infants and children, dietary fat delivers a major source of energy. The predominant fat source in breastmilk, infant formulas, and or the unrestricted child's diet is from long-chain triglycerides. In addition to serving as a concentrated source of calories, fatty acids play a role in cell signalling and gene expression, are a structural component of cell membranes, and participate in nervous tissue myelin production, all of which are important components of growth and development.[6] Additionally, both surgery and critical illness potentiate metabolic alterations by inducing the stress response, which results in the catabolism of endogenous stores of protein, carbohydrate, and fat to provide energy.[7]

The most common initial management strategy for chylothorax is to restrict long-chain triglycerides intake, altering the type of dietary fat, and thus promoting a diet enriched with medium-chain triglycerides. Fat absorption starts within the intestinal lumen. Fat globules known as micelles form after emulsification of dietary fats by bile acids. Long-chain triglycerides are converted to chylomicrons within the enterocytes, stimulating chyle production. Chyle is absorbed through lymphatic capillaries and in normal flow patterns travel through the lymphatic system via the thoracic duct to the bloodstream. Medium-chain triglycerides, consisting of triglycerides with saturated fatty acids of 8 to 12 carbon length, are absorbed directly into the portal venous circulation without micelle formation thus bypassing the lymphatic system and not contributing to chyle flow (Fig 1). Medium-chain triglycerides are suitable as an alternative fat source for the provision of additional calories in the setting of chylothorax.[8] The association between chylothorax resolution and amount and exposure to long-chain triglycerides has not been established. Additionally, when total fat is reduced or altered, other caloric sources will need to be optimised to ensure nutritional adequacy.

Neonates and infant diet management

Postoperative chylothorax is four times more likely to develop in neonates compared with older children[1] and can have detrimental effects on growth and clinical outcomes. Complete reduction of fat is not recommended as fat is crucial in provision of calories for sufficient growth and development. Optimizing nutrition delivery while reducing chylous drainage can present a challenge as the fat sources in human milk and traditional infant formulas are predominately long-chain triglycerides. Providing defatted fortified human milk or medium-chain triglycerides predominant infant formulas is safe and effective with adjunctive medical therapies in the treatment of chylothorax. Common practice includes trial of an medium-chain triglyceride-based infant formula or defatted human milk for 2-7 days and monitoring total chest tube drainage before considering parenteral nutrition.[9] Formulas commercially available for infants with chylothorax, specifically those with feeding intolerance or documented milk protein allergies, are limited. Often, off label use of toddler and adolescent formulas are required but should be done with close nutritional guidance to avoid nutrient deficiency. Recommended formulas for use in children <1 year of age with chylothorax are shown in Table 2.

Defatted human milk is preferred when available in an effort to avoid altering the gastrointestinal tolerance of breast milk. Fortunately, defatted human milk has been shown to be as safe and effective for the cessation of chylous drainage in comparison to traditional methods of stopping delivery of breastmilk and providing medium-chain triglycerides predominant infant formula.[10] Defatted human milk provides the known immunologic benefits of human milk to infants[11] and can promote sufficient growth with appropriate fortification.[12] Fat can be removed from human milk either through centrifugation, commercial cream separation, or natural separation techniques.[13,14] The most effective method for consistent fat removal is refrigerated centrifuge.[15] This process includes samples being placed in a refrigerated centrifuge at 2°C for 15 minutes at 3000 rpm. Once centrifuged, a transfer lid and syringe can be used to remove the defatted portion of breast milk. Secondary options are often considered given the financial implications with the regular use of a refrigerated centrifuge. Commercial cream separators have become a safe method for hospital and home use.[16] Natural separation is less reliable for consistent fat removal with the previously mentioned techniques being preferred.[15] Once fat is removed from human milk, fortification is required from medium-chain triglyceride-based formulas and or medium-chain triglyceride-rich calorie modular to optimise caloric density and delivery found in Table 2. Defatted human milk

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Table 2. Oral and enteral formulas for use in fat modified diets[3]

		Composition per 100 kcal of formula			
Age	Oral or Enteral Formula (%MCT/%LCT)	Total fat, g	MCT, g	LCT, g	LA, g
Infant	Mead Johnson™ Enfaport™ (83/17)	5.50	4.57	0.94	0.35
	Nestlé® Lipistart® (78/22)[5]	4.48	3.33	0.94	0.31
	Nutricia Monogen® (84/16)[1]	2.93	2.47	0.45	0.20
Pediatric	Mead Johnson™ Portagen® (87/13)[1, 2]	4.68	4.07	0.61	0.28
	Nestlé® Vivonex® Pediatric (69/31)[1]	2.90	2.00	0.90	0.49
Adult	Nestlé® Vivonex® RTF (41/59)[4]	1.16	0.48	0.68	0.36
	Nestlé® Vivonex® T.E.N. (0/100)[2, 4]	0.27	0.00	0.27	0.22
	Nestlé® Tolerex® (0/100)[2,4]	0.20	0.00	0.20	0.12
		Composition per container or prepared serving			
Fat amount	Oral Nutrition Formula	Protein, g	MCT, g	LCT, g	Energy, kcal
Minimal to no fat	Nestlé® Boost Breeze®	9	0	0	250
	Nestlé® Carnation Breakfast Essentials®	13	0	0-1	220
	Ensure® Clear Nutrition Drink	8	0	0	240
Standard fat	Nestlé® Boost® High Protein	20	0	6	240
	Ensure® High Protein Shake	16	0	2	160
		Composition per 1 mL or 1 g fat additive			
Fat Type	Modular Additives	Protein, g	MCT, g	LCT, g	Energy, kcal
MCT	Nestlé MCT Oil®	0.00	0.93	0.00	7.67
MCT & LCT	Nutricia Liquigen®[5]	0.00	0.45	0.01	4.50
MCT & LCT	MCTprocal™[5]	0.12	0.61	0.01	7.03

[1]Nutritionally complete but off label use in patients under 1 year of age.
[2]Nutritionally incomplete and requires trace mineral supplementation.
[3]Typical use for oral intake and not for tube feed.
[4]May be insufficient to meet essential fatty acid requirements.
[5]Total includes other types of fat not listed.

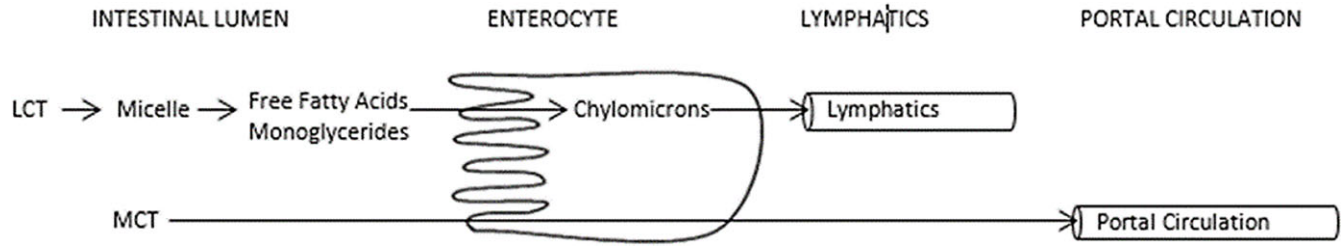

Figure 1. Medium Chain Triglyceride Absorption.

is 10-12 kcal/oz on average and is insufficient as a sole source of nutrition for infants.

Children and adolescent diet management

There is no consensus on what constitutes a fat-restricted diet in terms of total fat grams/day or caloric delivery derived from fats. The 2020 Dietary Guidelines for Macronutrients recommends for children ages 1-3 to consume 30-40% calories from fat, ages 4-18 to consume 25-35% calories from fat.[17] Fat modification for the purpose of chylothorax has been reported to be from <10 g total fat/day[18] to <30% of calories from fat/day.[19,20] Duration of effective therapy also varies among centres. The recommended duration of fat modification ranges between 10 days and 6 weeks for fat-modified diets.[21–24] Prolonged oral diets providing <10% calories from long-chain triglycerides for greater than 1-3 weeks put children at risk for essential fatty acid deficiency.[25] The amount of oral fat tolerated also depends on the age of the child, their caloric requirements, and the severity of disease. Nutrition optimisation during dietary fat restriction may include more meals, snacks, and oral supplements throughout the day to help reach nutrition goals. When transitioning to a fat-modified oral diet, a registered dietitian should be utilised to guide families on appropriate food choices. Suggested dietary modifications to

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Table 3. Suggested dietary modifications to supplement nutritional delivery during fat restriction

Food group	Consume MORE often (fat free or low-fat, ≤5 g/serving)	Avoid/limit foods with (>5 g/serving)
Fruits	Fresh and dried fruit Frozen and canned fruit Fruit juice Jelly, jam, or other fruit spreads Stage 1,2 baby foods, fruit based	Coconut cream (coconut water ok) Coconut meat
Vegetables	Raw vegetables Frozen and canned vegetables Cooked vegetables with no added fat/oil Vegetable juice and tomato juice Fat free tomato paste or sauce Pickles Salsa Stage 1, 2 baby food, vegetable based	Avocado Olives Cooked vegetables with added fat/oil Fried vegetables such as french fries, curly fries, tater tots, onion rings, tempura, or anything in batter Vegetables canned/jarred in oil such as giardiniera
Milk/Dairy/Milk-Free Alternatives	Milk (fat free or 1%) Yogurt (fat free or low fat) Cottage cheese (fat free or low fat) Cheese, cream cheese (fat free or low fat) Sour cream (fat free or low fat) Low fat ice cream & frozen yogurt/sherbet/sorbet Lower fat milk alternative (i.e. low fat soy milk versus regular soy milk)	Regular fat dairy (2% or whole) Creamers Coconut cream/milk Higher fat milk alternative (i.e. low fat soy milk versus regular soy milk)
Meat/Protein	Lunch meat (chicken & turkey are lower in fat) White meat poultry without skin Goat meat Lower fat fish (typically lighter in color) Tuna packed in water Mollusks Shrimp without head Crab without roe Low fat meat substitutes Beans and lentils cooked with no additional fat/oil Egg whites/egg substitutes (if yolk, use only one) Low-fat peanut butter powder	Hot dogs and sausages Beef, pork, mutton Poultry with skin & dark meat poultry Fried meats Fatty fish (typically darker in color) Soybeans/edamame/tofu Peanut butter/other nut butters Bean dips Nuts/seeds Whole eggs
Grains/starches	Breads/bagels/muffins without added fat/oil Tortilla Baked potato chips Air popped popcorn without added oil Cereals: - Rice Krispies® - Corn Flakes® - Frosted Flakes® - Shredded Wheat® - Special K® Crackers, rice cakes Rice, pasta, tubers	Bread/bagels/muffins with added fat/oil Tortilla chips Fried potato chips Seasoned microwave popcorn Granola Cereal w/ nuts Instant noodles Waffles/pancakes
Fats/Condiments/Other	Salad dressing (low fat) Mayonnaise (low fat, fat free) Salt, pepper, herbs, spices Honey, syrup, agave, sugar Broth/soups without milk/cream Ketchup, mustard, Relish	Salad dressing (regular) Mayonnaise (regular) Butter/margarine/lard/oil Gravy Dips Cream or cheese sauces Cream soups

Daily total fat should be considered. Check labels for serving size and number of fat grams.

supplement nutritional delivery during fat restriction are shown in Table 3.

The Fontan procedure carries the highest incidence of chylothorax and is associated with increased risk of mortality.[26] Proposed mechanisms for chylothorax as a consequence of the Fontan circulation include elevated systemic venous and hydrostatic capillary pressures, increased pulmonary vascular resistance, and low systemic vascular resistance, which leads to congestion of the lymphatic system causing increased production of lymphatic drainage.[5,26–30] To prevent chylothorax in this high-risk population, some have trialed *preoperative prophylactic* fat restriction. Sunstrom et al employed a diet with <30% calories from fat in conjunction with diuresis and fluid restriction in 14 patients in the immediate postoperative Fontan period until chest tube was removed at 6 days compared to previous average of 11 days prior to this intervention. Patients without chylous drainage resumed a

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Table 4. ASPEN parenteral nutrition administration.

	INITIATION		ADVANCE BY		GOALS	
	Preterm	Term	Preterm	Term	Preterm	Term
Infants (<1 y)						
Protein (g/kg/d)	1.5–3	1.5–3	1	1	3–4	2–3
CHO (mg/kg/min)	5–7	6–9	1–2.5% dextrose per day	1–2.5% dextrose per day	8–12 (Max 14–18)	12 (Max 14–18)
Fat (g/kg/d)	1–2	1–2	0.5–1	0.5–1	3-3.5 (Max 0.17 g/kg/hr)	3 (Max 0.15 g/kg/hr)
Children (1-10 y)						
Protein (g/kg/d)	1–2		1		1.5–3	
CHO (mg/kg/min)	10% dextrose		5% dextrose per day		8–10	
Fat (g/kg/d)	1–2		0.5–1		2–3	
Adolescents						
Protein (g/kg/d)	0.8–1.5		1		0.8–2.5	
CHO (mg/kg/min)	3.5 or 10% dextrose		1–2 or 5% dextrose per day		5–6	
Fat (g/kg/d)	1		1		1.2–5	

ASPEN Pediatric Nutrition Support Core Curriculum, 2nd Edition, 2015.[33]

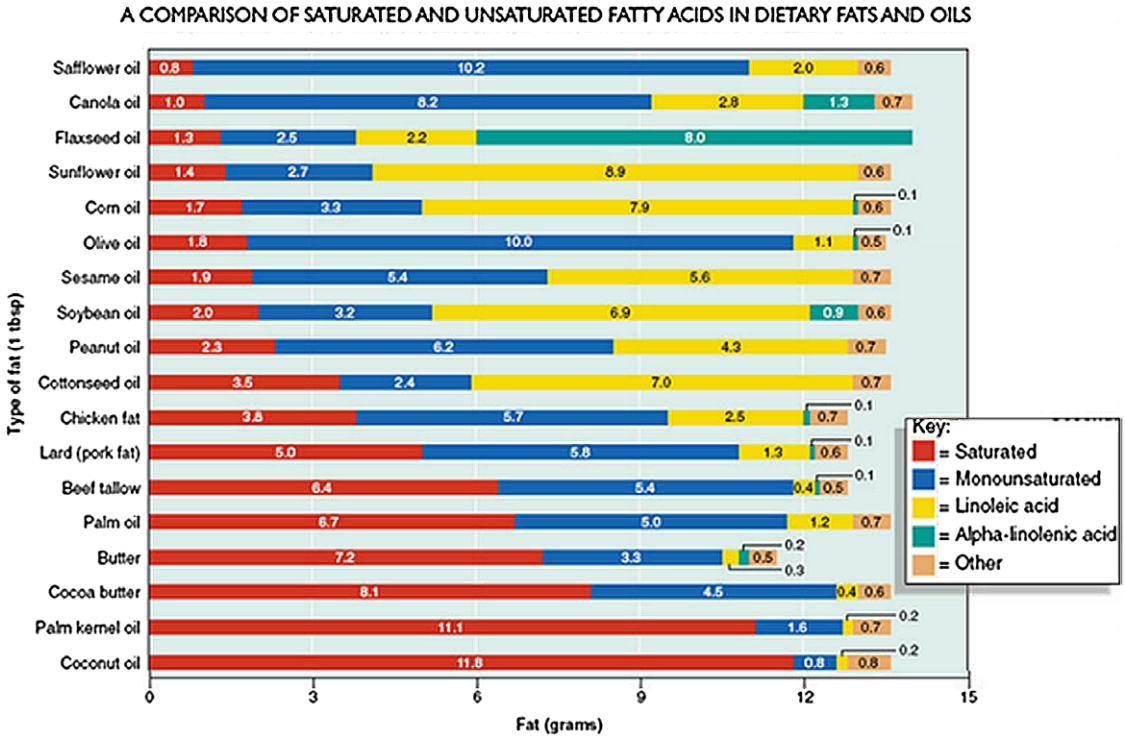

Figure 2. A Comparison of saturated and unsaturated fatty acids in dietary fats and oils.

normal diet post chest tube removal,while those with chylous drainage were discharged on fat restriction for 6 weeks from surgery. Pike and colleagues also provided a prophylactic fat restriction to 30% calories from fat in all Fontan patients for a full 6-week duration. Prophylactic fat restriction may provide the benefit of reducing lymphatic pressure and drainage, resulting in decreased chest tube days and hospital length of stay.[19,20]

The ability to reach goal caloric and protein requirements on a fat-modified diet may be challenging in the setting of a poor appetite. It is important to counsel patients and their families to be attentive to protein consumption given that low-fat foods are often low in protein. In some instances, oral or enteral supplementation may be required to obtain a low-fat, high-protein diet that is not otherwise achievable for those with

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Table 5. Suggested nutrition related biochemical monitoring for chylothorax.

Lab	Monitoring	Dosing Considerations	
Basic Metabolic Panel + Phosphorus	Daily initially, weekly monitoring once stable or at any point with concerns	Replace or supplement as needed based on levels	
LFTs	On Parenteral Nutrition		
Triglycerides	On Parenteral Nutrition Infants: >400 mg/dL, stop IL and recheck TG Children: >250 mg/dL reduce and recheck >400 mg/dL discontinue and recheck		
IGG	history of significant infections	Goal >500 mg/dL with risk of significant infections[43]	
Albumin	Daily initially, weekly monitoring once stable or at any point with concerns	Consider replacing by IV or parenteral	
Serum Zinc	Consider checking as early as two weeks or at any point if concerns for deficiency	**Enteral**	**Parenteral**
		2xRDA for age*	0.1 mg/kg/d (max 5 mg/d) (<5 yrs) 2.5–5 mg/d (>5 yrs)
Ceruloplasmin Copper, RBC		Ensure RDA for age*	20 mcg/kg/d (<1 yr) 200–500 mcg/day (>1 yr)
Serum Selenium		Ensure RDA- up to 2x RDA*	1–2 mcg/kg/day (<1 yr)
Retinol binding, Serum A		Ensure RDA then up to 1.5–2x RDA for age*	Provide MVI
Vitamin D 25-OH		10–50 mcg (400–2000 IU) daily of cholecalciferol*	Provide MVI
Vitamin E: serum, cholesterol and triglyceride levels**		Ensure RDA*; consider MVI	Provide MVI
Vitamin K: Plasma Prothrombin time/International Normalized Ratio (INR)		Ensure RDA	10 mcg/kg/d (<1 yr) 200 mcg/d (>1 yr)
Comprehensive Fatty Acid Profile including Triene: Tetraene (T:T), ALA, LA, and mead acid levels		0.2–1% of total calories from ALA 2–4% of total calories from LA	

*Do not exceed upper limit (UL).
**Lipid blood content suggested for accurate assessment of α-tocopherol, low levels during protein-energy malnutrition may falsely indicate deficiency.[44]

significantly reduced oral intake. Medium-chain triglycerides predominant or fat-restricted oral nutrition supplement should be considered to optimise nutritional intake as needed (see Table 2).

Parental nutrition therapies in high-volume chylothorax

If enteral fat modifications fail to reduce persistent high-volume chylothorax (>20 mL/kg/day), a period of nil per os with parenteral nutrition should be considered. There is a lack of consensus regarding the duration of nil per os. Prior studies reference an nil per os duration ranging from five to twenty-one days.[4,18,31] Fluid restriction when possible in conjunction with diuresis are common strategies for reducing central venous pressures, which may be exacerbating chylous drainage.[32] Hence, the caloric delivery via parenteral nutrition is often restricted particularly in the early postoperative period and even more so in those with chylothorax. Of note, intravenous lipid emulsions can be given while nil per os and are delivered directly into the bloodstream therefore bypassing the lymphatic system providing a valuable source of calories and essential fats.[33,34] The American Society for Parenteral and Enteral Nutrition provides nutrition care guidelines for critically ill children. Table 4 displays recommended parenteral nutrition initiation, advancement, and target macronutrient distribution for different age groups.

Special considerations while on fat-modified diet

Essential fatty acid deficiency

Medium-chain triglyceride-rich diets where >85% of the fat source is medium-chain triglycerides used for the treatment of chylothorax may not provide adequate amounts essential fatty acids.[9] Essential fatty acid deficiency results when there is insufficient dietary intake of linoleic acid and alpha-linolenic acid from a limitation of dietary fat or altered absorption of fat.[35] The human body is unable to synthesise linoleic acid (18:2 $\Delta^{9,12}$) and alpha-linolenic acid (18:3 $\Delta^{9,12,15}$) due to the lack of enzymes that can introduce double bonds beyond the Δ^9 site, therefore making them essential from the diet.[36] Essential fats are necessary for healthy cell membrane formation, cholesterol metabolism, blood clotting, as well as

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Table 6. Micronutrients commonly deficient in chylothorax and physical effects.

Nutrients		Role	Deficiency	Nutrition Focused Physical Exam Findings Associated with FSV and EFAD
Fat soluble Vitamins (FSV)	Vitamin A	Vision, gene expression, reproduction, embryonic development, growth, immune function	Poor dark adaptation, xeropthalmia, impaired immune function	Hair: Easily plucked hair, dull, dry, color changes Skin: seborrheic dermatitis (scaly, waxy, crusty plaques on scalp/lips); petechiae, purpura, hyperkeratosis, abnormal dryness, flakiness Nails: Beau's lines (transverse ridges, horizontal grooves) or transverse white lines Gums: gingivitis, swollen, bleeds easily, dental caries Skeletal: demineralization of bone, bowed legs, rickets, frontal bossing, bone tenderness Muscular: weakness, muscle cramps or pain Nervous: seizures, peripheral neuropathy w/ weakness Eyes: night blindness, Bitot's spots
	Vitamin D	Calcium and phosphorus regulation, bone growth, neuromuscular and immune function	Poor bone mineralization, increase risk of infection	
	Vitamin E	Antioxidant function	Peripheral neuropathy, ataxia, skeletal muscle weakness, increased erythrocyte fragility	
	Vitamin K	Hemostasis and bone metabolism	Hemorrhagic disease and defective blood clotting, reduced bone mineralization	
Minerals	Zinc	Component of enzymes involved in the maintenance of structural integrity of proteins and in the regulation of gene expression.	Poor growth, impaired appetite, alopecia, diarrhea, delayed sexual maturation, eye and skin lesions	Skin: Eczema, slow healing, scaliness Nails: Beau's lines (transverse ridges, horizontal grooves) Eyes: angular blepharitis(inflammation of eyelids) Taste: diminished taste, hypoguesia Gums: gingivitis, swollen, bleeds easily, dental caries
	Copper	Component of several metalloenzymes, which act as oxidases in the reduction of molecular oxygen	Normocytic hypochromic anemia, leucopenia, neutropenia, osteoporosis in children	Nervous: peripheral neuropathy with weakness; ataxia, decreased tendon reflexes Eyes: pale conjunctiva Hair: color change, depigmentation
	Selenium	Antioxidant, regulate thyroid hormone	Cardiomyopathy, Keshin-Beck disease	Hair: color change, depigmentation

Note: many micronutrients are affected by metabolic stress and may not be an accurate reflection of body stores.

proper brain and nervous system development.[37] Prevention of essential fatty acid deficiency can be achieved, respectively, with intakes of 2-4% of total calories from linoleic acid and 0.2-1% of calories from alpha-linolenic acid.[35,38]

In the presence of adequate enteral intake of essential fatty acids, tetraene products predominate in plasma. When the intakes of both linoleic acid and alpha-linolenic acid are low, triene formation is high and hence an elevated triene to tetraene (T:T) ratio. Mead acid (a triene) is produced at increased frequency in the absence of linoleic acid and alpha-linolenic acid due to enzymatic activity on non-essential fatty acids.[36] An *elevated T:T ratio >0.4* is considered diagnostic of essential fatty acids.[39] Biochemical evidence of essential fatty acids occurs earlier than the clinical signs and symptoms, which may include dry scaly skin, skin lesions, dry dull hair, brittle nails, hair loss, poor wound healing, and poor growth.[37,40] Screening for essential fatty acids via an essential fatty acid profile is recommended for patients receiving a fat-free diet for prolonged period (>3-4 wks), receive inadequate intravenous lipid emulsions/oil supplementation, and those who may lack adequate fat stores such as neonates and underweight children.[36,41] If the infant or child is starting to show biochemical or physical signs of essential fatty acids, linoleic acid, and alpha-linolenic acid-rich fat sources such as sunflower, safflower, corn, soybean, flaxseed, canola, walnut, and fish oils can be added while still maintaining an overall low total long-chain triglycerides nutrition plan, see comparison of oils in Figure 2.[42]

The addition of intravenous lipid emulsions could be considered to help meet the essential fatty acids requirement. For infants, linoleic acid must be provided at 3% or greater of total kilocalories to meet the essential fatty acids requirement. An intravenous lipid emulsions infusion of 2.5-5 mL/kg/day (0.5-1 g/kg/day) should be adequate to prevent essential fatty acid deficiency. In children, essential fatty acid deficiency can be avoided by providing 2.5 mL/kg/day (0.5 g/kg/day) of intravenous lipid emulsions. Intravenous lipid emulsions may be given daily at a minimal dose or throughout the week (i.e. Monday, Wednesday, and Friday).[34]

Biochemical monitoring

Biochemical monitoring, in addition to routine physical exams and tracking of somatic growth, is warranted for anyone receiving a fat-modified diet for >2 weeks. Large-volume output from chylothorax can cause high-protein losses, resulting in low albumin levels, and excessive losses of fat, electrolytes, immunoglobulins, and other minerals that may be protein bound, such as zinc, copper, and selenium.[35] These minerals should be closely monitored in patients receiving parenteral nutrition, specifically when delivery is reduced secondary to supply shortage.

Chyle is rich in lymphocytes and immunoglobulins that play a critical role in immune response and infection prevention. Hoskote and colleagues demonstrated a significant correlation between duration of chyle loss and degree of lymphopenia including

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an association between prolonged chylous drainage with a nadir on day 5 (range 2-6 days) in neonates and worse secondary immunodeficiency. In children, loss of immunoglobins was seen more in large volume losses and less with duration of loss.[43] A basic metabolic panel, phosphorus, albumin, and immunoglobulins level should be monitored. Supplementation and repletion of electrolytes, albumin, and/or immunoglobulins should be considered if abnormal, see Table 5 for recommended surveillance labs and timing.

Nutrition-focused physical exam monitoring

Nutrition-focused physical exam includes an examination of subcutaneous fat, muscle loss, oedema, and assessing for micronutrient-deficient specific clinical signs. A nutrition-focused physical exam should be performed on all patients at the time of chylothorax diagnosis and regularly throughout their treatment course to properly customise their nutrition interventions. As fat-soluble vitamin deficiencies, mineral deficiencies, and essential fatty acid deficiency are more common in this population, please see Table 6 for signs and symptoms of deficiency.

Discussion

To address the nutritional considerations unique to paediatric patients with chylothorax, we utilised the Chylothorax Work Group infrastructure to develop contemporary, clinically relevant, nutritional guidance based on expert experience, and literature review. The guidelines provide age-specific recommendations regarding delivery of fat-modified diets, management of parenteral nutrition, nutrition-focused physical exam, biochemical monitoring rationale, and recommendations. We suggest that optimal recovery from chylothorax will require clinicians to mitigate the myriad of nutritional insults associated with this complication. Within the Chylothorax Work Group, ongoing efforts are underway to better understand the nutritional influences of chylothorax and best practices when postoperative chylothorax occurs in children with children heart disease.

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