Nutrition management varies significantly depending on the defect, but remains crucial, throughout various care settings. Nutrition therapy is complicated by increased energy needs, reduced energy intake, and potential inefficient absorption and utilization of calories. Medical, surgical, and nutritional management is dictated by the type and severity of cardiac lesion. Furthermore, many nutrition-related complications and comorbidities, such as laryngeal dysfunction, necrotizing enterocolitis, protein losing enteropathy, chylothorax, and poor wound healing, can result from medical or surgical management of the defect. Quality of life for patient and family, as well as getting the child back on track for age-appropriate development, is always at the forefront of each care plan.

These guidelines seek to provide practical suggestions for the management of these infants and children with congenital heart disease and the comorbidities associated with them.

Feeding Guidelines

Post-operative fluid restriction and calorie requirements

- **Introduction**
  - Provision of nutrition after cardiac surgery requires careful consideration of the patient’s hemodynamic status, fluid balance, level of sedation and pain control, and return of gastrointestinal motility. Gastrointestinal perfusion may be drastically impacted in the post-operative period depending on the patient’s clinical status and the medications they are receiving. Additionally, total fluid intake may be limited to avoid fluid retention as the body builds an inflammatory response to the operation, which causes capillary leak and can lead to edema.

- **Critical Thinking Points**
  - Provision of nutrition is impacted by the fluid the patient is receiving from continuous and intermittent medications and flushes.
  - TPN should be withheld until it can provide approximately 30–40 ml/kg or more of total fluid intake.
  - Fluid intake may be restricted from 50 to 80 percent of maintenance fluid needs following cardiac surgery with cardiopulmonary bypass (Owens & Musa, 2009). As the patient recovers, medications are discontinued, and fluid balance is liberalized, the parenteral nutrition should be increased to a maintenance fluid goal in order to meet daily nutritional requirements.
• Diagnostic evaluation
  o Albumin and pre-albumin
  o Monitor daily weight when possible

• Treatment
  o Resting energy expenditure (REE) has been utilized to determine a patient’s minimum caloric requirement in the immediate post-operative period following cardiac surgery. The thermogenic effect of food is negated when receiving parenteral nutrition, meaning energy needs are approximately ten percent less than enteral nutrition requirements (Koletzko, et al., 2005). An infant should initially receive 55-60 kcal/kg/day during the acute post-operative phase in order to meet their basal energy requirements. During this time, it is understood that the patient is undergoing stress and as a result experiences increased proteolysis. They are often mechanically ventilated and on sedatives and/or neuromuscular blocking agents, and these factors reduce resting energy needs. As infants are recovering and becoming more active, it is recommended that the caloric content of their nutrition be advanced to a goal of 90-100 kcal/kg/day (Mirtallo, et al., 2004). The minimum caloric requirements and calorie goals for patients beyond the newborn period vary depending on age (see table below).

<table>
<thead>
<tr>
<th>Calorie Guidelines for Patients &gt;1 Month of Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial (kcal/kg/day)</td>
</tr>
<tr>
<td>-----------------------</td>
</tr>
<tr>
<td>35-45</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Maximum (kcal/kg/day)</th>
<th>1month-1yr</th>
<th>1yr-7yrs</th>
<th>7yrs-12yrs</th>
<th>&gt;12yrs</th>
</tr>
</thead>
<tbody>
<tr>
<td>85-105</td>
<td>75-90</td>
<td>50-75</td>
<td>30-50</td>
<td></td>
</tr>
</tbody>
</table>

(See Appendix A for Enteral Feeding Guidelines for Infants 0-4 months)

• Associated Complications
  o Potential for volume overload
  o Poor nutrition related to inability to tolerate the fluid volume necessary to administer enteral or parenteral nutrition
  o Poor wound healing associated with inadequate nutrition

Neonate
• Introduction of the problem
  Infants with congenital heart disease have impaired weight gain during the first several months of life. The type of cardiac lesion, hemodynamic compromise, and timing of surgical correction or palliation plays a role in growth failure in these infants.

• Critical thinking points
  o Fluid loss due to congestive heart failure
  o Energy intake required to sustain normal growth is higher
  o Gastroesophageal reflux (GER)

• Diagnostic evaluation of the problem
  o Nutritional markers (albumin, pre-albumin)
o Daily weight measurements
o Daily caloric measurements

- **Treatment**
  - TPN should be initiated perioperatively if enteral feeds are not indicated or as a supplement to minimize nutritional deficiencies
  - Increased caloric intake between 130 and 150 kcal/kg/d
  - Follow growth curve closely
  - Medical management of congestive heart failure (CHF), GER

- **Associated Complications**
  - Wound healing
  - Failure to thrive (FTT)

---

**Single Ventricle Patient**

- **Introduction of the problem**
  Infants with HLHS are known to be at risk for poor nutrition and growth failure. There is little published information regarding successful nutrition interventions for infants with single ventricle physiology. Feeding protocols have been shown to maximize the benefits and minimize the risks of enteral nutrition in the management of these critically ill patients. Although many feeding strategies have been proposed in neonates with HLHS, including standardized feeding protocols and preemptive gastrostomy tube (G-tube) placement, none have resulted in dramatic improvement in weight gain during the interstage period or become “standard care” to promote growth in this population. Because of this, there is significant center variation in feeding practice and growth outcomes in these high-risk infants.

- **Critical thinking points**
  - The Norwood hospitalization and early perioperative period is associated with a significant decrease in weight-for-age z-score (WAZ) and failure to thrive for most patients.
  - Factors contributing to growth failure in single ventricle infants include inadequate calorie intake, high metabolic demands, gastrointestinal pathology, and genetic and extracardiac abnormalities.
  - Despite the importance of weight gain, advancement to nutrition goals is often slow in the perioperative period. It is hampered by concerns for poor systemic output, the need for inotropic support, limitations of fluid intake, the risk of necrotizing enterocolitis (NEC), and frequent interruptions in nutrient delivery.
  - Post operatively there are concerns for cardiac and respiratory insufficiency, feeding intolerance, and laryngeal nerve injury.

- **Diagnostic evaluation of the problem**
  - Monitor nutritional markers (Albumin and pre-albumin)
  - Daily weight measurements
  - Monitor daily caloric intake
  - Speech Therapy evaluation of oral motor skills
  - Radiologic evaluation-modified barium swallow
  - Fiberoptic endoscopic evaluation of swallowing (FEES)
• **Treatment**
  o TPN should be initiated in all single ventricle patients preoperatively, and as soon as feasible postoperatively to minimize nutritional deficiencies
  o Increased caloric intake between 130 and 150 kcal/kg/d
  o Caloric density: generally 24-27 kcal/oz (may be limited by feeding intolerance)
  o Follow growth curve closely. Goal weight gain: average of 25-35 g/day
  o Medical management of GER

• **Associated Complications**
  o NEC
  o Vocal cord paralysis
  o Failure to thrive
  o Wound healing

• **Special Considerations**
  o Standardized feeding evaluation before discharge
  o Use of red flags
    ▪ Weight gain/loss (weight loss > 30 gm in 1 day
    ▪ Failure to gain 20 gm in 3 days, intake <100ml/kg/d)
  o Interstage home monitoring program (scales, saturation monitor) (See Interstage Monitoring Document)
<table>
<thead>
<tr>
<th>DAILY FOOD GUIDE FOR CHILDREN FOODS</th>
<th>1 TO 2 YEARS</th>
<th>2 TO 5 YEARS</th>
<th>5 TO 12 YEARS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Dairy</strong></td>
<td>2-3 servings: 8 fl oz. 2% Milk, 1 cup yogurt, 1/3 cup or 1 oz low fat cheese</td>
<td>2-3 servings: 8 fl oz. 1% or skim milk, 1 cup yogurt, 1/3 cup or 1 oz low fat cheese</td>
<td>3 or more servings: 8 fl oz. 1% or skim milk, 1 cup yogurt, 1/3 cup or 1 oz low fat cheese</td>
</tr>
<tr>
<td><strong>Grains/Breads (choose whole grain more often)</strong></td>
<td>1.5 - 3 servings: ½ cup pasta or rice or oatmeal, 1 cup cereal, 5 (1 oz) crackers, 1 tortilla, 1 slice of bread</td>
<td>4 - 5 servings: ½ cup pasta or rice or oatmeal, 1 cup cereal, 5 (1 oz) crackers, 1 tortilla, 1 slice of bread</td>
<td>5 servings or more</td>
</tr>
<tr>
<td><strong>Desserts</strong></td>
<td>fruit, frozen yogurt, sorbet, jello, custard, pudding, ice cream</td>
<td>fruit, frozen yogurt, sorbet, jello, custard, pudding, ice cream, plain cake or pastries</td>
<td>fruit, frozen yogurt, sorbet, jello, custard, pudding, ice cream, cake or pastries</td>
</tr>
<tr>
<td><strong>Fats</strong></td>
<td>As needed to make food palatable: oil, butter, salad dressings</td>
<td>As needed to make food palatable</td>
<td>As needed to make food palatable</td>
</tr>
<tr>
<td><strong>Fruits</strong> (Choose a rainbow of colors: oranges, yellows, reds, purples)</td>
<td>2 servings: ½ cup fresh/canned/frozen, 1 small piece of fruit, ¼ cup dried fruit, ½ cup 100% fruit juice</td>
<td>2-3 servings: ½ cup fresh/canned/frozen, 1 small piece of fruit, ¼ cup dried fruit, ½ cup 100% fruit juice</td>
<td>2-3 servings: ½ cup fresh/canned/frozen, 1 small piece of fruit, ¼ cup dried fruit, ½ cup 100% fruit juice</td>
</tr>
<tr>
<td><strong>Meat, Poultry, Fish, beans/lentils, tofu, eggs, nuts</strong></td>
<td>2-3 servings: 2-3 T. finely chopped meat, poultry or boned fish, ½ cup beans/lentils, 1 egg, 1 oz nuts, 1 T. nut butter, ½ cup tofu</td>
<td>3-4 servings: 1 oz lean meat, poultry, or fish, ½ cup beans, 1 egg, 1 oz nuts, 1 T. nut butter, ½ cup tofu</td>
<td>4-5 servings</td>
</tr>
<tr>
<td><strong>Vegetables (choose leafy greens, yellows, oranges, reds)</strong></td>
<td>2 servings: 1 cup cooked, chopped or mashed</td>
<td>2-3 servings: 1 cup cooked or tender raw vegetables</td>
<td>2-3 servings: 1 cup cooked or tender raw vegetables</td>
</tr>
<tr>
<td><strong>Miscellaneous</strong></td>
<td>Light seasoning: sugar, salt, jelly</td>
<td>Moderate: sugar, salt, honey</td>
<td>Moderate: sugar, salt, honey Any seasoning pepper, jams, jelly, catsup, mustard, pickle relish.</td>
</tr>
</tbody>
</table>
Guidelines to Manage Neonatal and Pediatric Feeding Concerns

Re-feeding Cardiac Patients Post-operatively

- Introduction
  - Assessment of feeding readiness after cardiac surgery requires an evaluation of multi-system readiness, including hemodynamic stability and gastrointestinal motility. Inability to tolerate feeds or limited gastrointestinal perfusion pre-operatively must be considered in the post-operative period, and may play a role in the time of initiation or rate of increase of feeds.

- Critical Thinking Points
  - Gastrointestinal perfusion may be limited pre-operatively in lesions of ductal dependent systemic blood flow such as interrupted aortic arch or coarctation of the aorta. Sudden increase in blood pressure below the obstruction results in necrotizing arteritis of the small arteries of the mesentery and small intestine. Patients may experience difficulties with reperfusion such as abdominal pain, tenderness, vomiting, ileus, melena, fever, and leukocytosis; this is referred to as postcoarctectomy syndrome (Nichols, et al., 2006).
  - Patients who have undergone surgical repair involving the aortic arch are at risk for damage to the recurrent laryngeal nerve and may experience vocal cord paresis or paralysis, resulting in impaired oral feeding skills.
  - Gastrointestinal motility may be impaired post-operatively due to effects from anesthesia and post-operative opioid use.

- Associated Complications
  - Post-coarctectomy syndrome- necrotizing arteritis of the small arteries of the mesentery and small intestine
    - Diagnostic evaluation
    - Symptomatic evaluation, abdominal x-ray, CBC
    - Treatment
    - Aggressive control of post-operative hypertension (Nichols, et al., 2006)
  - Vocal cord paralysis:
    - Diagnostic evaluation
    - Flexible endoscopic examination of vocal cord motion, evaluation by speech therapy, video swallow study
    - Treatment
    - Careful monitoring of feeding safety, and may require thickened liquids or direct gastric feeds
  - Ileus- intestinal dysmotility in the absence of a mechanical obstruction (Schnitzler, et al., 2008)
    - Diagnostic evaluation
    - Symptomatic evaluation: diminished or absent bowel sounds, failure to pass gas or stools, abdominal distention, vomiting, increased gastric residual volume
    - On abdominal x-ray may see increased air in the bowel or intestinal distention but may need abdominal CT to rule out mechanical obstruction
    - Prevention
    - Adequate fluid resuscitation
    - Rational use of vasopressors
• Weaning opioids
• Initiation of early enteral feeding
  • Treatment
    • Primary treatment is gastric decompression via NG tube to reduce vomiting (Schnitzler, et al., 2008)

Wound Healing
• Introduction
  o Post-operative nutrition plays a critical role in wound healing. Congenital heart disease patients may be malnourished pre-operatively, and may also have higher metabolic demands.

• Critical Thinking Points
  o If the cardiac patient has open wounds or delayed sternal closure, this presents more nutritional challenges. The patient is more susceptible to the loss of lean body mass due to higher baseline requirements.
  o The provision of adequate protein is the single most important nutritional intervention in ill or post-operative children. Protein is required to maximize protein synthesis and preserve skeletal muscle protein mass, as well as to facilitate wound healing and the inflammatory response (Rodriguez-Key & Alonzi, 2007).

• Diagnostic evaluation
  o There are no specific guidelines for neonates or children in regards to nutritional treatment for pressure ulcers.
  o There are laboratory tests that are recommended to be obtained at baseline when following a wound:
    ▪ Albumin, Prealbumin
    ▪ Metabolic panel
    ▪ Total blood count
  o Testing for vitamin absorption and deficiencies in vitamins A, C, D, B2, calcium, zinc, and copper is suggested (Rodriguez-Key & Alonzi, 2007).
  o Daily weight
  o Careful evaluation of the sternal wound

• Treatment
  o Supplemental Vitamin C, Folic Acid, Zinc, and arginine (Shulman & Phillips, 2003; Skillman & Wischmeyer, 2008)

• Associated Complications
  o Wound dehiscence

Post-operative Chylothorax
• Introduction
  o Chylothorax is a post-operative complication that results in leakage of lymphatic fluid into the pleural space. Chylothorax may occur due to surgical disruption of the thoracic duct or one of its main tributaries. Additionally, it may be a result of increased pressure within the intrathoracic lymph system (Chan, et al., 2005).
• **Critical Thinking Points**
  - Incidence of post-operative chylothorax is higher following heart transplantation and Fontan procedures (Chan, et al., 2005).
  - Post-operative chylothorax is associated with prolonged length of stay (Chan, et al., 2005).

• **Diagnostic evaluation**
  - Chest x-ray evidence of pleural effusion
  - Chest ultrasound to evaluate for pleural effusion
  - If chest tube is in place and the patient has been receiving enteral nutrition, the drainage has a creamy appearance because it contains chylomicrons and long-chain triglycerides. However, in the post-operative period the patient is often fasting and the drainage may continue to appear serosanguinous.
  - Laboratory evaluation of the pleural fluid can be performed to suggest chylothorax, but it is not confirmative. Parameters include a triglyceride level of >1.2 mmol/L and a total cell number of >1000 cells/microliter, predominantly lymphocytes (Zuckerberg & Lefton-Greif, 2006).

• **Treatment** *(See chylothorax care map flowsheet from Chan, et al., 2005 -Appendix B)*
  - Initial management may involve chest tube placement for drainage of large effusions.
  - Transition the enteral formula to one that does not contain long-chain triglycerides, such as Enfaport or Vivonex.
  - If a change in formula is not effective, enteric rest and provision of total parenteral nutrition may be required (Chan, et al., 2005; Zuckerberg & Lefton-Greif, 2006).
  - Octreotide may be prescribed for chylothorax that is refractory to dietary changes. It acts directly on splanchnic vascular receptors to reduce chylomicron synthesis and transport into the lymphatic duct, which decreases lymphatic flow rate and limits triglyceride loss (Zuckerberg & Lefton-Greif, 2006). This therapy has been shown to have variable efficacy (Chan, et al., 2005).
  - Patients who fail both dietary and medical management will require surgical interventions, such as ligature of the thoracic duct (most common), pleurodesis with talc/tetracycline or fibrin glue, video-assisted thoracoscopic surgical (VATS) identification and clipping of the site of duct leakage, or pleuroperitoneal shunting (Zuckerberg & Lefton-Greif, 2006).

• **Associated Complications**
  - Large chyloous effusions are associated with immune compromise due to loss of T lymphocytes, nutritional deficits due to loss of chylomicrons, and cardiovascular derangements due to loss of electrolytes into the pleural space (Zuckerberg & Lefton-Greif, 2006).

• **Special Considerations**
  - Clinical improvement is defined as a decrease in effusion drainage to <10 ml/kg/day (Zuckerberg & Lefton-Greif, 2006).
  - Refractory chylothoraces may require a careful hemodynamic evaluation, such as cardiac catheterization, to evaluate for etiology of systemic venous hypertension, such as systemic venous thrombosis, pulmonary artery stenosis, elevated pulmonary vascular resistance, or elevated right ventricular end diastolic pressure (Zuckerberg & Lefton-Greif, 2006).
Modifying breast milk by decanting fat, neonate receives skimmed breast milk – study in progress at Doernbecher Children’s Hospital

Protein Losing Enteropathy

- **Introduction of the Problem:** (Rychik 2007, Braamskamp 2010)
  Protein losing enteropathy (PLE) is one of the known complications of the Fontan procedure. It usually results from the increased pressure on the right side of the heart. Within 10 years after a Fontan procedure, 13.4% of patients may develop this complication. Mortality in patients who develop PLE after a Fontan procedure may be as high as 56%.

- **Critical Thinking Points:**
  - Loss of absorption of protein
  - Significant loss of immunoglobulins and lymphocytes
  - Increased risk for infection
  - Compromised absorption of fat-soluble vitamins

- **Diagnostic Evaluation:**
  - Diagnosis mainly based upon clinical symptoms
    - Edema
    - Diarrhea
  - Formerly a diagnosis of exclusion, now with 3 current tests
    - Nuclear test with ingested, tagged proteins identified in stool
    - MRI to identify lymphatic abnormalities
  - Easiest and most commonly laboratory used: stool sample to measure the endogenous protein, Alpha 1-Antitrypsin (A1-AT).
    - A1-AT is not present in the diet, has a similar molecular weight to albumin, and is normally not actively secreted, absorbed, or digested.
    - It is not excreted in urine, so even stool sample collected in a diaper will provide accurate results.

- **Treatments:** (Lin 2006, Personnel communication, Tortorich 2014)
  - Multiple treatments with a varied degree of success
    - Diet therapy
    - Diuretics
    - Afterload reduction
    - Corticosteroids
    - Heparin
    - Protein infusions
    - Surgical revision of baffle with fenestration
    - Heart Transplantation
  - Dietary modifications in patients with PLE related to lymphatic pathology
    - Encourage low fat, high protein diet
    - Maintain adequate energy from fat source
    - Decrease lymphatic circulation with limitation of fat
• Absorption of long-chain triglycerides (LCT) from gut stimulates lymphatic flow
• In normal diets medium-chain triglycerides (MCT) and LCT occur in the same foods (i.e. coconut oil contains 60% MCT, but still has 40% LCT)
  ▪ If fat supplement needed: MCT oil most common supplement
  ▪ Add supplements of fat-soluble vitamins (A,D,E,K) and minerals
  ▪ AquADEKs Pediatric Liquid, ADEKs Chewable
  ▪ May need to consider sodium restriction
  o Tips to teach to patients/parents/caregivers (Tortorich 2014)
    ▪ Encourage the child to have:
      • A good protein source with each meal (3/day) and snack (2-3/day) good sources are:
        o Low/no-fat dairy
        o Very lean neat/poultry/seafood
        o Egg whites
        o Low-fat tofu
        o Beans
      • More common to need a fat free protein powder than to add MCT oil
    ▪ Good overall oral supplement:
      • Skim milk with instant breakfast powder
      • Pediasure SideKicks juice
      • Ensure clear
    ▪ Parent Handout (See Appendix C, Low Fat Diet)
References:

Post – operative patient


Neonate - Single ventricle patient


**Chylothorax**


**Protein Losing Enteropathy**


**Additional resource:**
Nutrition Care Manual
An excellent nutritional resource, however you must have a membership to access. Most hospitals have dietitians or hospital departments that have a membership.

The Nutrition Care Manual is an Internet-based professional resource that is updated annually and used internationally. Purchased by annual subscription, the Nutrition Care Manual is available online and easily linked to an Intranet system, enabling all staff to quickly access it without need for user names and passwords.

The Nutrition Care Manual is consistent with the Centers for Medicare & Medicaid Services' Interpretive Guidelines for the Hospital Conditions of Participation. It also meets the Joint Commission's Hospital Accreditation Standards 2009.

- Whether in a clinical or an academic setting, users can rely on the following features to find the most up-to-date and accurate nutrition information.
- The Academy's online Nutrition Care Manual includes Disease-specific nutrition assessment, diagnosis, intervention, monitoring and evaluation.
- Printable client handouts, including foods recommended list, foods not recommended list and a sample one-day menu.
- Ten calculators that compute BMI/weight range, resting energy expenditure, diabetic exchanges and more. Customization tools such as formulary database of nutrition solutions, electronic sticky notes and electronic highlighting. An extensive formulary database that is updated semiannually and easily customizable to any facility.
Appendix A

Doernbecher PICU Enteral Feeding Guidelines – Infant (0-4 months) Cardiac Surgery Patients

- Place enteral feeding tube within 12 hours of admission.
  - NJ feeding is desirable, NG feeds will be considered in patients with low aspiration risk.
  - Daily assessment for starting enteral feedings.
  - Enteral feeds are appropriate with good end organ perfusion, stable respiratory status, no evidence/low risk of NEC.
  - If feeding is contraindicated, continue or resume TPN. POD 0-1 initiate low electrolyte, concentrated TPN.
  - Daily weight on all patients.

Contraindications to Feeding
- Intestinal obstruction/ileus/NEC
- Impending procedure
- Significant respiratory distress
- High dose vasopressors (Dopamine >3 or Epi >0.03)
- Escalating support
- Elevated lactate (>2)

Feeding Selection
- Expressed mother’s breast milk (EMM) is preferred
  - If breast milk is not available start 20 cal/oz formula-Good Start or Similac Total Comfort or home formula
  - See chylothorax algorithm if chest tube output appears milky, change formula to Enfaport

Feeding Initiation and Advancement

<table>
<thead>
<tr>
<th>Weight</th>
<th>Start</th>
<th>&quot;1 ml/kg/hr&quot;</th>
<th>&quot;1 ml/kg/hr&quot;</th>
<th>&quot;120 ml/kg/day or 5 ml/kg/hr&quot;</th>
<th>TPN Wean</th>
</tr>
</thead>
<tbody>
<tr>
<td>2.2-2.5 kg</td>
<td>2 ml/hr</td>
<td>2 ml/hr q 4 hrs</td>
<td>10-12 ml/hr</td>
<td>4 ml/hr</td>
<td></td>
</tr>
<tr>
<td>2.5-3 kg</td>
<td>2 ml/hr</td>
<td>2 ml/hr q 4 hrs</td>
<td>10-12 ml/hr</td>
<td>4 ml/hr</td>
<td></td>
</tr>
<tr>
<td>3.1-3.5 kg</td>
<td>3 ml/hr</td>
<td>3 ml/hr q 4 hrs</td>
<td>16-18 ml/hr</td>
<td>6 ml/hr</td>
<td></td>
</tr>
<tr>
<td>3.6-4 kg</td>
<td>3 ml/hr</td>
<td>3 ml/hr q 4 hrs</td>
<td>18-20 ml/hr</td>
<td>6 ml/hr</td>
<td></td>
</tr>
<tr>
<td>4.1-4.5 kg</td>
<td>4 ml/hr</td>
<td>4 ml/hr q 4 hrs</td>
<td>20-22 ml/hr</td>
<td>8 ml/hr</td>
<td></td>
</tr>
<tr>
<td>4.6-5 kg</td>
<td>4 ml/hr</td>
<td>4 ml/hr q 4 hrs</td>
<td>23-25 ml/hr</td>
<td>8 ml/hr</td>
<td></td>
</tr>
<tr>
<td>5.1-5.5 kg</td>
<td>5 ml/hr</td>
<td>5 ml/hr q 4 hrs</td>
<td>26-27 ml/hr</td>
<td>10 ml/hr</td>
<td></td>
</tr>
<tr>
<td>5.6-6 kg</td>
<td>5 ml/hr</td>
<td>5 ml/hr q 4 hrs</td>
<td>28-30 ml/hr</td>
<td>10 ml/hr</td>
<td></td>
</tr>
</tbody>
</table>

- Adjustments in the goal rate will be determined by the dietitian and team.

Advancement Criteria: Stable respiratory and hemodynamic status, stable abdominal girths, no emesis or diarrhea.

Intolerance: (abdominal distention, increasing abdominal girth, or emesis) consider holding feeds for 2 hours or returning to previous volume/concentration. Notify medical team if persistently not tolerating feeds.

Fortification: Once tolerating goal feeding rate for 24 hours, fortify feeds to 24 cal/oz (formula or EMM + formula).

Most infants will need to then further increase the feeding rate ("145 ml/kg/day or 6 ml/kg/hr) or concentration (27-30 cal/oz).

Aspiration Risk Factors: History of aspiration or feeding intolerance, cleft lip/ palate, vocal cord abnormality, aortic arch repair

NI to NG: Consider transitioning from continuous NI to NG feeds in patients tolerating goal feeds & have a low risk of aspiration.

Exubation
- If patient passes ERT, hold NG feeds 6 hrs prior to extubation or hold NI feeds 2 hrs prior to extubation.
- Evaluate feeding status 2 hrs after extubation and restart feeds if respiratory status is stable.

Oral and Bolus Feeds
- Transition to oral and/or bolus feeds when tolerating full NG feeds and on stable respiratory support <3L HFNC.
- Consult SLP to assess suck, swallow, and help maximize oral intake.
- ENT consult to assess vocal cords for all aortic arch repairs (HLHS, CoA, IAA) and in patients with a hoarse cry.
- Initiate breast feeding in addition to tube feeding, will gradually adjust tube feedings as infant improves at nursing.

Bolus Feeds: Hold continuous feeds for 2 hours. Divide 24 hour continuous feeds into bolus feeds. Start with q 2 hr feeds x 4 feeds (ie- 20 ml/hr = 40 ml q 2 hrs), then increase to q 2.5 hr feeds x 4 feeds (ie- 20 ml/hr = 50 ml q 2.5 hrs), and finally increase to q 3 hr feeds (ie- 20 ml/hr = 60 ml q 3 hrs). Give boluses over 1 hour initially and gradually advance to 30 minutes.

If safe per SLP feed PO for up to 20 minutes then NG gavage the remainder.

Once taking 75% or more at each feeding, remove NG for trial of all PO feeds. Evaluate intake daily for need to fortify further or replace NG tube.

Adapted from Lucile Packard Children’s Hospital Stanford, Sandy Staveski.
Appendix B

Chylothorax Decision Tree – The Hospital for Sick Kids – Toronto, Ontario,

PERSISTENT CT DRAINAGE
(=6ML/KG/DAY) on PO/N/4
OR
PRESENCE OF MILKY CT DRAINAGE

YES
- Chyle (+)
- Chyle (-) plus Lymph >80%

NO

Phased thoracic imaging (ultrasound, ECHO to rule out thoracic duct ligation)

Diagnostic imaging (ultrasound, ECHO to rule out thoracic duct ligation)

If thrombus, Rx with heparin & tPA

PHASE 1
MCT diet/ minimal fat diet x5 days

YES
- NO

NO

If thoracic duct ligation considered early

For early reconstruction, notify CVS to consider early thoracic duct ligation

PHASE 2
TPN/PO

YES
- Start TPN x5 days
- Vascular access consult for PICC
- JP/CT off suction if decreasing drainage

NO
- Return to phase 2 for min 7 days

Phase 3

YES
- Start Prednisone p.o.
- (2mg/kg/day divided bid) duration of 5-7 days
- JP/CT off suction if decreasing drainage
- Return to MCT/minimal fat diet
- d/c TPN when adequate enteral caloric intake reached

NO

Phase 4

YES
- Start Ocreotide (0.5 - 4mg/kg/day IV continuous infusion) x5 days
- Ocreotide 500mg/kg/dose IV q6H if limited IV access (increase q24h by 50mg/kg/dose to max 40mg/kg/dose x 5 days
- d/c Prednisone
- JP/CT off suction if decreasing drainage
- Continue min fat/MCT diet

NO

Phase 5

YES
- Surgical Intervention (eg. Thoracic duct ligation etc)
- m-PTRP pathway post-op at Phase 1
- Consider pleuropertitoneal shunt if CT continues to drain

NO

PHASE 3

YES
- Wean Prednisone as per HSC Formulary
- Continue min fat diet/MCT diet

NO

Return to Phase 2 for min 7 days

CHAITHORAX DECISION TREE
- THE HOSPITAL FOR SICK KIDS, TORONTO, ONTARIO, CANADA


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## Low Fat Diet

<table>
<thead>
<tr>
<th>Food Group</th>
<th>Foods to Include</th>
<th>Foods to Avoid</th>
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</thead>
<tbody>
<tr>
<td><strong>Grains</strong></td>
<td>Pasta (un filled), bread, yeast rolls, oatmeal, tortillas, potatoes, pretzels, rice, quinoa, barley, baked chips, low fat- popcorn, cereal, crackers, graham crackers, muffins, waffles, pancakes</td>
<td>Fried potatoes, potato chips, filled pasta, regular- pastries, muffins, cereal, granola, waffles, pancakes, corn bread, sweet rolls, crackers</td>
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<tr>
<td><strong>Vegetables</strong></td>
<td>Fresh, frozen, canned without added fat</td>
<td>Avocado, olives, buttered/creamed/fried veggies</td>
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<tr>
<td><strong>Fruits</strong></td>
<td>Fresh, frozen, canned without added fat</td>
<td>Coconut</td>
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<tr>
<td><strong>Dairy</strong></td>
<td>Skim/fat free or 1% milk including flavored milk, low fat cheese (&lt;3 gm fat/oz), non fat yogurt, non fat frozen yogurt, non fat cottage cheese, fat free pudding, fat free sour cream</td>
<td>2% or whole milk including flavored milk, heavy cream, half n half, regular ice cream, milk shakes, custard, regular cheese, regular yogurt, soups made with whole milk</td>
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<tr>
<td><strong>Protein</strong></td>
<td>Baked, boiled, grilled, broiled lean meat, fish, poultry, low fat deli meat, egg substitutes/egg whites, tofu, legumes, water packed tuna, low fat hot dogs, beans</td>
<td>Ham, sausage, bacon, frankfurters, fish canned in oil, fried meat, poultry, fish, sardines, spiced or pickled meat, duck, goose, egg yolks, nuts, nut butters</td>
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<tr>
<td><strong>Fats/Oils/Sugar</strong></td>
<td>Fat free mayo and salad dressing, fat free whipped cream, popsicles, sherbet, sorbet, jam/jelly, honey, jello, jelly beans, hard candies, angel food cake, fruit ice</td>
<td>Butter, margarine, oil, lard, regular mayo and salad dressing, gravy, desserts made with whole milk or cream, cake, pies, pastries</td>
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<tr>
<td><strong>Miscellaneous</strong></td>
<td>Herbs, lemon juice, pepper, salt, spices, vinegar, extracts, condiments: catsup, chili sauce, mustard, pickle relish, soy sauce</td>
<td>Coconut, nuts, gravy, sauces</td>
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</tbody>
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Low Fat Diet

Description: A diet that contains less then 10-15% of calories from fat. Generally this is 25-35 grams of fat per day for older children and adults, 10-20 grams of fat per day for younger children.

Tips for Keeping Foods Low-fat

- Use lean meat or tofu with lots of vegetables in stir fry
- Add flavor by cooking with lemon juice, herbs, spices, garlic
- Use evaporated non fat milk to thicken sauces instead of whole milk
- Use low fat cheese or feta cheese in pasta dishes like lasagna
- Choose lean cuts of beef and pork, like those labeled loin or round
- Remove visible skin and fat from meats before and after cooking
- Bake, broil, grill, roast, stew, or stir-fry lean meats
- Choose lean ground beef (7% fat or less) and drain off fat when cooking
- Sauté or bake white fish and season with lemon and herbs
- Use egg whites (or egg substitute) for eating, baking, and cooking

Meal and Snack Ideas

<table>
<thead>
<tr>
<th>Breakfast ideas</th>
<th>Cold cereal with skim milk, fruit, juice</th>
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<tbody>
<tr>
<td></td>
<td>Fat free yogurt, toast with jam, juice</td>
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<tr>
<td></td>
<td>Egg whites or egg substitute, fruit, toast with jam, skim milk</td>
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<tr>
<td>Snacks</td>
<td>Fruit, low-fat granola bar, fat free yogurt, smoothie</td>
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<tr>
<td>Lunch ideas</td>
<td>Sandwich with lean turkey and fat free mayo, fruit, skim milk</td>
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<tr>
<td></td>
<td>Quesadilla with low-fat cheese, salsa, fat free sour cream</td>
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<td></td>
<td>Soup, low-fat crackers, fruit, carrots, skim milk</td>
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<tr>
<td>Snacks</td>
<td>Pretzels, baked chips, string cheese, low-fat granola bar, popsicle, low-fat beef jerky</td>
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<tr>
<td>Dinner ideas</td>
<td>Baked chicken strips, mashed potatoes, vegetable, skim milk</td>
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<tr>
<td></td>
<td>Mac-n-cheese (box type made without added butter, skim milk), vegetable, skim milk</td>
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<td></td>
<td>Pasta with marinara sauce, turkey meat balls, vegetable, skim milk</td>
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<tr>
<td>Snacks</td>
<td>Popsicle, sorbet, sherbet, angel food cake, fat free yogurt, smoothie</td>
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</tbody>
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Doernbecher Children's Hospital, Portland, Oregon; provided by Alisa Tortorich RD, CSP, CNSC, LD