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Nourishing Little Hearts: Nutritional Implications for Congenital Heart Defects



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Congenital heart defects (CHD) result from abnormal formation of the walls or valves of the heart or associated vessels in the fetal period. Medical, surgical, and nutritional management is dictated by the type and severity of cardiac lesion. Patients with CHD present unique nutritional challenges, as they generally have higher energy and nutrient needs, yet many factors negatively impact their abilities to consume, absorb, or utilize substrate. Furthermore, many nutrition-related complications and comorbidities, such as laryngeal dysfunction, necrotizing enterocolitis, acute renal failure, and chylothorax, can result from medical or surgical management of the defect (adding insult to injury). This review seeks to provide practical suggestions for the management of these infants and children in the pre-operative neonatal, post-operative critical care, acute care/step-down, and post-discharge settings.

INTRODUCTION

Definition, Classification, Cause, and Diagnosis

Congenital heart defects (CHD) are characterized by anomalies in the structure of the heart and its related valves and vessels; these defects are present at birth and typically present in infancy. Defects range from clinically irrelevant to hemodynamically and physiologically significant. Defects can be classified as either cyanotic (right-to-left shunting) or acyanotic (left-to-right shunting). In many cases, the cause of

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the defect in a particular infant is unknown; from an epidemiological standpoint, maternal smoking during pregnancy, genetics, and chromosomal abnormalities have been cited as factors contributing to CHD (1). Common defects presenting a nutrition challenge are presented in Table 1. CHD are diagnosed prenatally via fetal echocardiography (if the mother has a high-risk pregnancy or there are known risk factors for CHD in the fetus), or postnatally after the presentation of a heart murmur, cyanosis, or failure to thrive.

Incidence and Prevalence

The incidence of CHD has been reported to be 1% in the United States (2) and 0.8% in European countries (3). While CHD are relatively rare, they are the most

Table 1.
Common congenital heart defects that can have nutritional consequences (1,26,41)

<i>Defect name</i>	<i>Defining features</i>
<i>Acyanotic defects</i>	
Ventricular septal defect (VSD)	Abnormal opening in the septum between the two ventricles
Atrial septal defect (ASD)	Abnormal opening in the septum between the two atria
Patent ductus arteriosus (PDA)	Fetal ductus arteriosus fails to close, resulting in shunting of oxygenated blood from the aorta to the pulmonary arteries
Atrioventricular septal defect (AVSD)	Failure of the septum between the two atria and two ventricles to form properly
<i>Cyanotic defects</i>	
Tetralogy of Fallot (TOF)	A constellation of 4 defects: a large VSD, pulmonary stenosis, right ventricular hypertrophy, and an overriding aorta
Pulmonary stenosis (PS)	Narrowing of the pulmonary valve
Pulmonary atresia (PA)	Abnormal formation or absence of the pulmonary valve
Tricuspid atresia (TA)	Abnormal formation or absence of the tricuspid valve
Aortic stenosis (AS)	Narrowing of the aortic valve
Hypoplastic left heart syndrome (HLHS)	Underdevelopment of the left side of the heart, including the mitral and aortic valves, left ventricle, and aorta
Interrupted aortic arch (IAA)	Absence or discontinuation of part of the aortic arch
Coarctation of the aorta (CoA)	Narrowing of the aorta
Transposition of the great arteries (TGA)	Positions of the pulmonary artery and aorta are reversed
Total anomalous pulmonary venous return (TAPVR)	Pulmonary veins incorrectly connect to the right atrium instead of the left atrium
Double outlet right ventricle (DORV)	Both the pulmonary artery and the aorta arise from the right ventricle
Double inlet left ventricle (DILV)	Only the left ventricle is properly developed (underdeveloped right ventricle); both the left and right atria empty into the left ventricle; may also have TGA and VSD
Truncus arteriosus	The pulmonary artery and aorta are combined to form one single great vessel or trunk that override the left and right ventricles

common birth defect responsible for mortality in the neonatal period (2). In fact, CHD are responsible for more neonatal deaths than all other birth defects combined. The most prevalent defects recorded in the United States between 2004 and 2006 were atrioventricular septal defects (AVSD) (4.71/10,000 births) and Tetralogy of Fallot (TOF) (3.97/10,000 births) (4). Surgical techniques have greatly improved the rate of survival since the first heart surgeries were performed in the 1950s; as such, currently, about one million American adults are living with CHD (1).

CHD can occur in both term and premature infants and are more common in children with certain genetic syndromes. For instance, approximately 45–50% of children with trisomy 21 (Down syndrome) have a CHD, namely a type of septal defect. Other genetic defects commonly presenting with CHD include Noonan syndrome, Williams syndrome, and Turner syndrome (5).

Concomitant medical issues such as prematurity or genetic disorders can complicate nutritional management of the CHD patient, particularly when relating to estimating energy needs and promoting oral feeding.

MEDICAL AND SURGICAL MANAGEMENT

Some infants have insignificant defects that may resolve spontaneously early in life or manifest later in adolescence or adulthood, thereby avoiding the need for treatment in childhood. However, most children with CHD require some form of medical and/or surgical management, which can vary widely based on the type and severity of the defect(s), gestational age of the infant and clinical condition or acuity. Additionally, medical management may be drastically different depending on the phase of care the infant is currently in: pre-operative

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phase, the immediate post-operative/critical care phase, the step-down/acute-care phase, or post-discharge care. While this review focuses on the nutrition management of the pediatric CHD patient, it is important to consider the effects of medical and surgical management on nutrition status and the nutrition care plan that may follow.

Medical Management

CHD patients may require oxygen to maintain oxygen saturations in a range that is acceptable for their particular defect. If the oxygen is to be delivered via nasal cannula, this may impact the availability of a nare for the placement of a nasogastric tube (NGT) for enteral nutrition (EN). Of note, there are one-prong pediatric nasal cannulas available for this reason. Patients may be receiving a plethora of medications that have nutrition-related side effects; these medication doses and types will change frequently throughout the various phases of care. Common nutrition-related medications are presented in Table 2.

Surgical Management

Defects may be corrected or palliated by a cardiothoracic surgeon in the operating room or an interventional cardiologist at cardiac catheterization. The type of defect dictates the intervention required, although there are various surgical approaches to palliation or correction of a given defect. Some of the simpler defects, such as ventricular septal defects (VSDs) or atrial septal defects (ASDs), may be corrected using a patch to repair the defect. The more structurally and nutritionally challenging defects, however, require palliation using mul-

tiple surgical procedures spaced out over many months or years. For instance, infants with hypoplastic left heart syndrome (HLHS) or other single-ventricle physiology typically require a three-stage procedure to re-route oxygenated and unoxygenated blood flow so that the properly-formed side of the heart (typically the right side) is made to be the main pumping chamber. The first stage is completed in the neonatal period, and the third stage is typically done around 3 years of age.

Table 2.
Common CHD medications that can have nutrition-related side effects (42,43)

<i>Medication Name</i>	<i>Nutrition-Related Side Effect or Concerns</i>
Antiarrhythmic	
Amiodarone	Nausea, vomiting, constipation, abdominal pain, anorexia
Lidocaine	Nausea, vomiting
Procainamide	Diarrhea, nausea, vomiting, abdominal pain, anorexia
Anticoagulant	
Warfarin (Coumadin)	Need consistent intake of vitamin K; diarrhea, nausea, GI pain/cramps, anorexia
Diuretics	
Bumetanide (Bumex)	GI cramps, nausea, vomiting, electrolyte abnormalities
Chlorothiazide (Diuril)	Anorexia, nausea, vomiting, electrolyte abnormalities
Furosemide (Lasix)	Electrolyte imbalance, hypokalemia, anorexia
Spirololactone (Aldactone)	Increased serum potassium (potassium-sparing), diarrhea, GI cramps, nausea, vomiting, anorexia, gastritis
Inotropes	
Digitalis (Digoxin)	Nausea, vomiting, anorexia, feeding intolerance, electrolyte imbalance
Dopamine	Nausea, vomiting
Dobutamine	Nausea, vomiting
Epinephrine	Increased myocardial oxygen consumption, nausea, weakness
Pain Control/Sedation/Paralytics	
Fentanyl	Nausea, vomiting
Midazolam (Versed)	Decreased energy needs, nausea, vomiting
Cisatracurium, pancuronium, vecuronium (paralytics)	Decreased energy needs
Vasodilators	
Nitroglycerin	Nausea, vomiting, abdominal pain, dry mouth
Sildenafil	Nausea, diarrhea
Other Medications	
Prostaglandin E1 (Alprostadiil)	Diarrhea

Table 3.
Criteria for determination of malnutrition in the pediatric CHD patient* (20,27)

- Weight for age <3rd percentile
- Height/length for age <3rd percentile
- Weight for length <3rd percentile
- Reduction in >2 percentiles for weight for age, length/height for age, and/or weight for length

*Measurements should be plotted on growth charts to evaluate percentiles and growth trends. The Centers for Disease Control and Prevention (CDC) recently recommended that measurements for infants <24 months of age be plotted on the 2006 modified WHO growth curves, but that measurements for children >24 months of age be plotted on the CDC curves (44). Utilize specialized growth charts for infants with trisomy 21.

Infants requiring a multi-stage palliation or repair are at greater nutrition risk than infants with simpler defects or those not requiring any sort of intervention. This is because persistence of the anatomical defect affects energy needs and intake, and surgeries and hospitalizations may affect nutrition status, intake, and needs. Furthermore, nutrition risk is elevated in these infants, as the neonatal, infant, and toddler stages of life can be critical periods to establish proper growth, development, and feeding behaviors.

For some infants with complex defects, heart transplantation may be the first-line intervention for management of the CHD in the neonatal period or may be required as the result of a failed palliative or corrective procedure later in infancy or adolescence (6). Approximately 25% of pediatric heart transplants are done in infants less than one year of age, and CHD are the impetus for almost two-thirds of transplants done in pediatric patients (7).

MALNUTRITION: PREVALENCE, ETIOLOGY, AND CONSEQUENCES

Success in managing the CHD patient is no longer solely dependent on survival after surgery. Since surgical interventions have a higher success and survival rate, the focus of CHD management has turned to maximizing growth, development, and quality of life and minimizing malnutrition. Malnutrition in this pop-
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Table 4.
Etiologies of malnutrition in the pediatric CHD patient (12–15,27)

Inadequate intake

- Anorexia from cardiac status or as a side effect of medications
- Fatigue while feeding
- Swallowing abnormalities, including uncoordinated suck, swallow, breathe; abnormal oral transit time; choking; gagging
- Oral aversion
- Neurological dysfunction as a result of prematurity or operative complications
- Laryngeal dysfunction
- Frequent vomiting or evidence of GERD
- Early satiety and decreased gastric volume due to hepatomegaly and/or ascites in CHF
- Tachypnea
- Fluid restriction
- Frequent periods of NPO for medical procedures/issues in the hospital
- Recurrent respiratory infections
- Psychosocial issues, including suboptimal feeding patterns/hunger cues and interactions with caregivers; parental feeding anxiety, particularly if artificial feeding methods are required; and financial limitations

Increased energy needs

- Chronic metabolic stress response to CHF
- Post-operative metabolic stress
- Relatively increased lean body mass in relation to fat mass in the malnourished patient
- Tachypnea
- Tachycardia
- Cardiac hypertrophy
- Polycythemia to compensate for chronic hypoxia
- Increased sympathetic nervous system activity
- Infections, fever, sepsis

Inefficient nutrient utilization/absorption

- Vomiting
- Reduced splanchnic blood flow
- Poor gastric emptying and altered gut motility
- Edema of the small bowel wall/mucosa (as a consequence of right sided heart failure), leading to malabsorption of nutrients
- Excessive nutrient loss, including protein-losing enteropathy or steatorrhea
- Excessive trace nutrient losses, i.e. calcium and potassium
- Gut mucosal atrophy leading to malabsorption in patients with significant pre-existing malnutrition
- IGF-1 deficiency

Abbreviations: GERD = gastroesophageal reflux disease; CHF = congestive heart failure; IGF-1 = insulin-like growth factor 1

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Table 5.
Nutrition assessment parameters for the pediatric CHD patient (12,21,27,44–46)

Food/Nutrition-Related History

- Food/beverage intake (older children)
- Breast milk/infant formula intake (infants)
- Enteral/parenteral nutrition intake
- Energy, fluid, macronutrient, and micronutrient intake (from all sources, including oral intake, enteral or parenteral nutrition)
- Medications with nutrition-related side effects
- Vitamin/mineral supplement use
- Feeding behaviors, including aversion, refusal, and fatigue
- Breastfeeding success/problems
- Food/nutrition program participation
- Adherence to prior nutrition education/instructions, such as proper formula mixing
- Physical activity (older children)

Anthropometric Measurements

- Height/length for age
- Weight for age (estimated euvolemic weight)
- Weight for length (infants <24 months old) or body mass index (children >24 months old)
- Ideal weight for height/length
- Head circumference (<3 years old)
- Growth pattern and percentile ranks
- Triceps skinfold measurements
- Abdominal girth

Biochemical Data and Related Tests

- Serum electrolytes (sodium, potassium, chloride, bicarbonate, calcium, magnesium, phosphorus)
- Blood glucose
- Blood urea nitrogen and creatinine measurements
- Hepatic profile
- Nutritional anemia profile
- Prealbumin in conjunction with CRP when prealbumin begins to increase as CRP decreases, may signify shift from catabolism to anabolism)
- Modified barium swallow study or flexible endoscopic evaluation of swallow (FEES) study

Nutrition-Focused Physical Findings

- Overall Appearance
- Cardiopulmonary system—increased work of breathing, wet lung sounds
- Extremities, muscles, and bones—loss of subcutaneous fat, muscle wasting, peripheral edema, peripheral cyanosis, muscle tone
- Digestive system—compromised swallow function, difficulty coordinating suck, swallow, breathe, abdominal distention or pain, poor appetite, ascites, nausea/vomiting/reflux, diarrhea
- Eyes and head—sunken eyes, sunken fontanelle, hair loss, temporal wasting
- Neurological changes
- Skin—dermatitis, dry/scaly, poor wound healing
- Vital signs—blood pressure, heart rate, respiratory rate, oxygen saturations, temperature

Patient History

- Type and severity of cardiac lesion
- Medical and surgical history, including prematurity
- Family history
- Socioeconomic factors, including family and living situation
- Genetic potential for growth

ulation can be difficult to define; suggested criteria are presented in Table 3.

At birth, children with certain types of CHD, including TOF, HLHS, VSD, and AVSD, tend to have lower birth weights, even when corrected for gestational age (8). Even if a child is born with a weight that is appropriate for

gestational age, failure to thrive typically becomes apparent in the neonatal period if hemodynamically significant CHD is present (9). In fact, estimates of malnutrition in the unrepaired CHD infant range from 50% to 90% (10,11).

The degree and type of malnutrition may be related to characteristics of the CHD, including pres-

Table 6.
Determination of estimated nutrition needs for the infant with CHD (9, 13, 14, 26, 47)

	<i>Critical care</i>	<i>Step-down/acute care</i>
Energy	Determine by indirect calorimetry, if available Provide REE (~55–60 kcal/kg) in the first 3–5 days after surgery or until CRP is <2 mg/dl	120–150 kcal/kg; 140–200 kcal/kg for catch-up growth Catch-up growth equation (kcal/kg): $\frac{\text{Kcal/kg for weight age} \times \text{ideal body weight}}{\text{current weight}}$
Protein	Term: 3–3.5 g/kg Preterm or LBW: 3–4 g/kg	
Fluid	As per critical care fluid restriction (generally 50–80% MIVF with liberalization after medication wean and sternal closure)	<3 kg: 120 mL/kg >3 kg: 100 mL/kg Consider +10–15% to compensate for increased losses with tachypnea, diarrhea, emesis, and diuresis
Micronutrients	Potassium: 2–5 mEq/kg Sodium: at least 2–3 mEq/kg even if sodium restriction is required Supplement iron and vitamin D in breastfed infants and those with low formula intakes	

Abbreviations: REE = resting energy expenditure; CRP = c-reactive protein; LBW = low birth weight; MIVF = maintenance intravenous fluids

ence of cyanosis, congestive heart failure (CHF), or pulmonary hypertension (PHTN). Infants with cyanosis tend to have similar alterations in both weight and length, while infants with acyanotic lesions tend to have more pronounced decreases in weight gain velocity as compared to length (12,13). It would stand to reason that the severity of growth impairment would be directly correlated to the severity of the cardiac lesion, but, instead, it seems the duration of cyanosis (years) is more important in determining severity of growth deficits (14). Descriptions of growth status in acyanotic and cyanotic lesions have changed over the past 25 years, as surgical intervention now occurs earlier in infancy for children with cyanotic lesions. As such, more recent studies have shown an emergence of more pronounced growth retardation and wasting in acyanotic infants (8). CHF and PHTN, which can occur in both cyanotic and acyanotic CHD, further complicate growth status and impairment by affecting factors that contribute to malnutrition (13,14).

Etiology of Malnutrition

The etiology of malnutrition in the pediatric CHD patient can generally be grouped into three categories: inadequate intake, inefficient absorption and utilization, and/or increased energy needs. In any one patient,

any or all of these factors may be present, which can make it difficult to delineate the optimal nutrition and medical therapy. Infants with genetic defects, prematurity, intrauterine growth retardation, or other congenital medical issues may have difficulties in any of these three etiological areas for reasons unrelated to the CHD, which further complicate management. Specific factors known to cause malnutrition in the CHD population are presented in Table 4. Of note, there is no consistent evidence to support the idea that most infants with CHD have alterations in absorption or energy utilization; however, this etiology may apply to the individual patient (15).

Consequences of Malnutrition

In the short term, infants and children with pre-operative malnutrition have reduced ability to fight infection or to heal optimally from surgical wounds (14,16,17). Intensive care unit (ICU) and total hospital length of stay are both prolonged in post-operative CHD infants with poorer nutrition status (17–19).

In the long term, malnutrition in infancy can produce suboptimal growth and physical and cognitive development later in childhood and adolescence. Some children experience significant catch-up growth after corrective or palliative surgeries; however, it seems

Table 7.
Nutrition-related goals for the pediatric CHD patient
(9,14,26,27)

Pre-operative neonatal period

- Provide adequate nutrition (preferably via EN) to meet the patient's needs until surgery

Post-operative critical care

- Initiate nutrition support as soon as possible to prevent development/worsening of malnutrition, minimize the loss of LBM, and to support functioning of vital organs
- Avoid refeeding syndrome in the infant with significant malnutrition
- Avoid overfeeding, which can cause difficulty in weaning from the ventilator
- Reduce unnecessary cessation of EN

Step-down/acute care

- Provide adequate nutrition to meet needs and correct nutrient or electrolyte deficiency
- Transition from PN to 100% EN
- Transition EN to oral or combination oral/supplemental EN; develop EN or oral schedule appropriate for home
- Demonstrate age-appropriate or catch-up weight gain and growth
- Parents/caregivers demonstrate competency in delivering appropriate nutrition prior to discharge

Post-discharge/home setting

- Demonstrate age-appropriate or catch-up weight gain and growth
- Demonstrate age-appropriate feeding behaviors

Abbreviations: EN = enteral nutrition; LBM = lean body mass; PN = parenteral nutrition

that infants operated on earlier have a better chance of increasing weight and length growth velocity to achieve maximal genetic potential. Other children may have persistent lesions or other genetic and congenital anomalies that affect growth after surgery or may have missed the crucial window of opportunity for appropriate growth. Children missing this critical window of opportunity may have reduced cell numbers (including adipose, muscle, and bone cells) that continue to yield small body size and weight. Furthermore, reduced cell numbers can also translate to reduced brain tissue (20). Brain weight can be reduced by as much as 30% in children with the most severe CHD (16). Reduced cell numbers can not only result in poor growth and delayed skeletal maturation, but also in poor motor skill development, including oral motor skills (14).

Delayed puberty is possible in children with impaired linear growth, as puberty does not typically begin until the child reaches the height of a normal 12–13-year old (13,21). Finally, children with CHD, particularly those under 3 years of age and with a cyanotic lesion, tend to have lower than normal intelligence scores, which could be a result of alterations in brain weight yielding reduced cognitive functioning (21).

NUTRITION MANAGEMENT

The focus of nutrition management of the pediatric CHD patient varies between the various phases of care. Appropriate nutrition care of the pediatric CHD patient first begins with thorough nutrition assessment; suggested parameters are included in Table 5. Determination of nutrition needs is outlined in Table 6, and Table 7 includes some examples of nutrition goals for CHD patients.

Pre-operative Feeding

Pre-operative EN in the CHD neonate, particularly one with potential poor gut perfusion due to ductus arteriosus-dependent circulation or low cardiac output, is a highly controversial topic, given the increased risk of necrotizing enterocolitis (NEC) (22). In fact, about 50% of physicians and nurse practitioners surveyed report *never* allowing EN (oral or tube feeding) pre-operatively in the single ventricle patient (23). Despite variability in practice, the current evidence indicates that it is reasonable to attempt EN in this population as long as the infant does not require high-dose pressors or does not show clinical or radiographic signs of NEC (22,24,25). Pre-operative intermittent oral and/or NGT feeding has been successful in multiple studies, with up to 95% of infants successfully receiving some sort of EN, up to 75% receiving at least 100 mL/kg/day, and about 1/3 receiving full EN prior to surgery. For patients who cannot be enterally fed, parenteral nutrition (PN) is the remaining option.

Post-operative ICU Feeding

In the immediate post-operative period, hemodynamic stability is the foremost concern in the ICU. Once the

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Table 8.
Guidelines for the initiation, advancement, and monitoring of parenteral nutrition therapy in the pediatric CHD patient (48)

Component	Initiation	Advancement	Goal	Monitoring
Fluid/volume	Per ICU team	Per ICU team to goal to meet fluid needs	Maintenance fluid needs or restriction as per ICU	Monitor for signs/symptoms of fluid overload or dehydration
Dextrose	10–15% depending on blood glucose levels; DDR* >4 mg/kg/min for infants	2–5% per day	To meet calorie needs	Blood glucose levels
Protein (Trophamine + L-cysteine for children <2 years of age)	2–3 g/kg/day or as limited by PN volume	1 g/kg/day	Infants: 3–4 g/kg/day Children: 2 g/kg/day	BUN
Lipids	1–2 g/kg/day	1 g/kg/day	Infants: 2–3 g/kg/day Children: 1–2 g/kg/day	Serum triglyceride levels
Sodium	Infants: 2–6 mEq/kg/day Children: 2–4 meq/100 mL			Serum electrolytes; Adjust by 0.5–1 mEq/kg/day or 0.5–1 meq/100 mL based on laboratory values
Potassium	Infants: 2–6 mEq/kg/day Children: 2–3 meq/100 mL			
Chloride	Infants: 2–8 mEq/kg/day Children: 2–3 meq/100 mL			
Calcium	Infants: 2–5 mEq/kg/day Children: 0.5–2 meq/100 mL			
Phosphorus	Infants: 1–1.5 mMol/kg/day Children: 0.75–1.2 mmol/100 mL			
Magnesium	Infants: 0.25–2 mEq/kg/day Children: 0.3–0.5 meq/100 mL			
Pediatric Multivitamin Solution	2 mL/kg/day with a maximum of 5 mL/day			
Pediatric Trace Element Solution	0.2 mL/kg/day with a maximum of 1 mL/day Consider reduction of dose with acute renal failure			
Selenium	1–2 mcg/kg/day if not already included in trace element solution			
Zinc	Additional 100 mc/kg for infants <2.5 kg or 50 mcg/kg for infants with open chest			
Famotidine	1 mg/kg/day			
Heparin	1 unit/mL PN volume			

Abbreviations: ICU = intensive care unit; DDR = dextrose delivery rate; BUN = blood urea nitrogen; PN = parenteral nutrition
 *DDR is calculated by the following formula: $DDR (mg/kg/min) = (\% \text{ dextrose} \times PN \text{ rate}) / (\text{weight in kg} \times 6)$
 Adapted with permission from the *UVA Children's Hospital Pediatric Nutrition Support Handbook (48)*

infant is stable hemodynamically, attention can be paid to other important factors, such as nutrition. If an infant is clinically stable soon after extubation, such as in a simple VSD repair, oral feeding may be possible relatively soon after return from surgery. However, in

many cases of complicated CHD, infants will require PN and/or EN. For many reasons, including cost, infection risk, and similarity to normal human physiology, EN is preferred over PN. However, in some cases, hemodynamic issues may preclude EN. PN is indi-

Table 9.
Advantages and disadvantages of various feeding routes in pediatric CHD patients (14,46)

<i>Feeding Route</i>	<i>Advantage</i>	<i>Disadvantage</i>
Nasogastric (NGT)	Short term Minimally invasive, can be placed at the bedside Physiologic feeding into the stomach	Difficulty breathing with tube in one nare Difficulty swallowing with tube in esophagus Contribute to esophagitis and worsen reflux Concern for use in the home setting (appropriate placement, inadvertent removal, vasovagal response)
Small bowel (nasoduodenal or nasojejunal)	Short term Helpful if reflux, vomiting, or delayed gastric emptying are present May allow a larger volume of feeds to be delivered over time	More difficult to place than NGT Difficulty breathing with tube in one nare Difficulty swallowing with tube in esophagus Contribute to esophagitis and worsen reflux Does not necessarily prevent tracheal aspiration of gastric contents Generally not appropriate for use in the home setting Requires continuous/drip feedings
Orogastric	Helpful for infants who cannot tolerate NGT due to nose-breathing or small nares Can be placed at the bedside	Difficulty securing the tube Need to remove when commencing oral feeds Generally not appropriate for use in the home setting
Gastrostomy	Less difficult to secure No interference with breathing or swallowing May be safer in the home environment than other tubes	Requires an additional procedure to place; usually is done in the operating room

cated when the expected time to begin and achieve adequate EN is more than 3–5 days (9,14). PN is also indicated when the gastrointestinal tract cannot be used for other reasons besides cardiac complications, such as other congenital anomalies involving the gut. PN delivery in the ICU is often limited because of total fluid restriction and medication drips required to maintain cardiac function, sedation, and pain control. However, the clinician should collaborate with ICU staff to maximize fluid available for nutrition delivery by concentrating medication drips as much as possible. Table 8 includes general guidelines for the initiation and advancement of PN for the CHD patient.

EN, even at trophic levels (0.5–1 mL/kg/hr), should be commenced as soon as medically feasible in order to reduce the risk of gut atrophy and loss of the intestinal mucosal barrier, which increases the risk of bacterial translocation and bacteremia (14,26,27). EN can be delivered via NGT or small bowel tube. An NGT is generally preferred, but in some scenarios a small bowel feeding tube may be more appropriate

(see Table 9). If a patient is expected to require a feeding tube for longer than 6–8 weeks or upon hospital discharge, a gastrostomy or jejunostomy tube should be considered. Table 9 outlines advantages and disadvantages of each feeding route.

EN is generally delivered using a continuous drip in the ICU due to the small hourly volume load, which presumably decreases complications such as aspiration and osmotic diarrhea (9,27). Continuous EN may allow for increased nutrient intake and assimilation because it minimizes myocardial oxygen consumption (14,27–29). For infants, the optimal feeding fluid is expressed breast milk (EBM), when available. If EBM is not available, standard infant formula is a reasonable substitute. There is no evidence to support the use of an oral rehydration solution as the first feeding fluid in the post-operative period. In fact, at this author's institution, the primary infant feeding fluid in the ICU setting is breast milk or standard infant formula fortified to 24 kcal/oz. General guidelines for the initiation and advancement of EN are presented in

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Table 10.
Indications for use of various infant and enteral formulas in the pediatric CHD patient

Category	Brand Names	Indication
Term Infant formulas		
Milk-based	Enfamil [®] Premium ¹ Similac [®] Advance ^{®2}	Standard infant feeding
Milk-based calorically dense	Enfamil Lipil [®] 24 ¹ Similac Expert Care [®] 24 ²	Infants with high energy needs or fluid restriction
Soy-based	Enfamil [®] Prosobee ^{®1} Gerber [®] Good Start [®] Soy ³ Similac [®] Soy Isomil ^{®2}	Lactose intolerance, galactosemia, some cases of milk-protein intolerance
Partially hydrolyzed	Enfamil [®] Gentlease ^{®1} Gerber [®] Good Start ^{®3}	Fussiness and gas
Prethickened	Enfamil A.R. ^{®1} Similac Sensitive for Spit-Up ^{™2}	Gastroesophageal reflux
Lactose free	Similac Sensitive ^{®2}	Lactose intolerance
Premature Infant Formulas		
Premature	Enfamil Premature [®] 24 ¹ Enfamil Premature [®] 24 High Pro ¹ Similac [®] Special Care [®] 24 ² Similac [®] Special Care [®] 24 High Pro ²	Prematurity
Premature high calorie	Similac [®] Special Care [®] 30 ²	High calorie needs or fluid restriction in prematurity; can be used as a breast milk fortifier
Premature transitional	Enfamil [®] Enfacare ^{®1} Similac Expert Care [®] Neosure ^{®2}	For infants >1800 grams
Specialized Infant Formulas		
Extensively hydrolyzed	Nutramigen ^{®1} Pregestimil ^{®1} Similac Expert Care [™] Alimentum ^{®2}	Allergy to intact proteins; fat malabsorption (may contain 30–55% of fat as MCT)
Amino acid	Elecare Infant ^{®2} Neocate Infant [®] DHA/ARA ⁴ Nutramigen [®] AA ^{™1}	Severe intact protein allergy (contain varying degrees of fat as MCT)
Fat-modified	Enfaport ^{™1} Monogen ^{®4}	Fat malabsorption or chylothorax (up to 85% of fat as MCT)
Renal	Similac [®] PM 60/40 ²	Reduced minerals/electrolytes
Pediatric Formulas (1–10 years of age)		
Standard	Boost [®] Kid Essentials 1.0 ⁵ Compleat [®] Pediatric ⁵ Nutren Junior ^{®5} Pediasure [®] Enteral ²	Standard pediatric feeding
Calorically dense	Boost [®] Kid Essentials 1.5 ⁵ Pediasure [®] 1.5 ²	High energy needs or fluid restriction
Protein hydrolysate	Pediasure Peptide [®] (1.0 and 1.5) ² Pepdite Jr. ^{®4} Peptamen Jr. [®] (1.0 and 1.5) ⁵	Gastrointestinal intolerance of standard formula
Amino acid	Elecare Junior ^{®2} Neocate Junior ^{®4} Vivonex [®] Pediatric ⁵	Severe gastrointestinal intolerance or protein allergy
Renal	Renastart ^{™6}	Reduced electrolytes

Abbreviations: MCT = medium chain triglycerides

Formula manufacturers: ¹Mead Johnson Nutritionals, Evansville, IN (www.mjn.com; 1-800-BABY-123); ²Abbott Nutrition, Columbus, OH (www.abbottnutrition.com; 1-800-227-5767); ³Nestle Infant Nutrition, Fremont, MI (www.medical.gerber.com; 1-800-628-BABY); ⁴Nutricia North America, Gaithersburg, MD (www.shsna.com; 1-800-365-7354); ⁵Nestle Nutrition, Florham Park, NJ (www.nestle-nutrition.com; 1-800-422-ASK2); ⁶Vitaflor USA, Alexandria, VA (www.vitaflorusa.com; 1-888-VITAFLO)

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Table 11.
Infant formula and expressed breast milk (EBM) concentration recipes

Kcal/oz	Term and specialized infant powders ^a	Term or specialized liquid concentrate	Premature transitional	Neocate Infant [®] DHA/ARA ¹	Enfaport [®] liquid ²	EBM
20	Standard dilution 1 sc per 2 oz water	Standard dilution 1 oz conc per 1 oz water	N/A	Standard dilution 1 sc per 1 oz water	1 oz liquid per 0.5 oz water	N/A
22	5 oz: 2.5 sc + 4.5 oz water 16 oz: 8 sc + 14.5 oz water 24 oz: 12 sc + 22 oz water	5.5 oz: 3 oz conc + 2.5 oz water 16 oz: 9 oz conc + 7 oz water 23.5 oz: 13 oz can conc + 10.5 oz	Standard dilution: 1 sc per 2 oz water	4 oz: 4.5 sc + 3.5 oz water 16 oz: 18 sc + 14 oz water 24 oz: 26.5 sc + 21 oz water	1.5 oz liquid per 0.5 oz water	½ sc term/ specialized powder + 8 oz EBM
24	4.5 oz: 2.5 sc + 4 oz water 16.5 oz: 9 sc + 15 oz water 26 oz: 14 sc + 23 oz water	4 oz: 2.5 oz conc + 1.5 oz water 17 oz: 10 oz conc + 7 oz water 22 oz: 13 oz can conc + 9 oz water	4 oz: 2 sc + 3.5 oz water 16 oz: 8.5 sc + 14 oz water 24 oz: 12 sc + 21.5 oz water	4 oz: 5 sc + 3.5 oz water 16 oz: 19 sc + 14 oz water 24 oz: 29 sc + 21 oz water	2 oz liquid per 0.5 oz water	½ sc term/ specialized powder + 4 oz EBM
27	8 oz: 5 scoops + 7 oz water 16 oz: 10 scoops + 14 oz water 24 oz: 15 scoops + 21 oz water	4.5 oz: 3 oz conc + 1.5 oz water 12 oz: 8 oz conc + 4 oz water 19 oz: 13 oz can conc + 6 oz water	8 oz: 4.5 sc + 7 oz water 16 oz: 9 sc + 14 oz water 24 oz: 13.5 sc + 21.5 oz water	4 oz: 5.5 sc + 3.5 oz water 16 oz: 21.5 sc + 13.5 oz water 24 oz: 32.5 sc + 20.5 oz water	4 oz liquid per 0.5 oz water	1 sc term/ specialized powder + 5 oz EBM
30	6 oz: 4 sc + 5 oz water 16 oz: 11 sc + 14 oz water 24 oz: 16.5 sc + 21 oz water	4 oz: 3 oz conc + 1 oz water 12 oz: 9 oz conc + 3 oz water 17 oz: 13 oz can conc + 4 oz water	4 oz: 2.5 sc + 3.5 oz water 16 oz: 10 sc + 14 oz water 24 oz: 15 sc + 21 oz water	4 oz: 6.5 sc + 3.5 oz water 16 oz: 24 sc + 13.5 oz water 24 oz: 36 sc + 20 oz water	Liquid is 30 kcal/oz as standard	1 sc term/ specialized powder + 4 oz EBM

Abbreviations: sc = scoops; conc = concentrate; EBM = expressed breast milk

^aIncludes standard and specialized infant powders as presented in Table 10, with the exception of Neocate Infant DHA/ARA Formula manufacturers:

¹Nutricia North America, Gaithersburg, MD (www.shsna.com; 1-800-365-7354)

²Mead Johnson Nutritionals, Evansville, IN (www.mjn.com; 1-800-BABY-123)

Figure 1, and Table 10 presents indications for different infant and enteral formulas. In many cases, because of fluid restriction and increased nutrient needs, infants with CHD will require caloric densities of >20 kcal/oz. Table 11 presents common strategies to increase the caloric density of infant formulas and breast milk.

Many pediatric cardiology programs have developed, or are in the process of developing, algorithms to guide initiation and advancement of EN in the post-

operative period. Several papers support this strategy as being effective in reducing PN requirement, days needed to reach goal EN, and gastrointestinal morbidity for ICU patients (30,31), and CHD patients in particular (32–35). There are fundamental differences in all of these successful algorithms; perhaps more important than the specific feeding parameters are the systematic initiation and advancement of feedings, consistent criteria that define feeding intolerance, and strategies to manage obstacles to EN.

Table 12.
Nutrient content of common baby foods (50,51)

<i>Food</i>	<i>Amount</i>	<i>Calories</i>	<i>Protein (g)</i>	<i>Fat (g)</i>
Hawaiian delight dessert, stage 2	4 oz	110	2	0
Vanilla custard with bananas dessert, stg 2	4 oz	110	3	1.5
Whole milk yogurt, fruit-flavored	4 oz	110	4	4
Chicken and gravy, stage 2	4 oz	100	8	6
Avocado, mashed	2 oz	90	1.2	8.4
Banana, plum, grape, stage 2	4 oz	90	1	0
Bananas, stage 2	4 oz	90	0.5	0.1
Sweet potatoes and turkey dinner, stage 2	4 oz	90	2	1
Banana and mixed berries, stage 2	4 oz	80	1	0
Chicken noodle dinner, stage 2	4 oz	80	3	2.5
Green beans, stage 2	4 oz	80	2	0
Ham and gravy, stage 2	4 oz	80	8	4
Peach cobbler dessert, stage 2	4 oz	80	1	0
Pear strawberry granola, stage 2	4 oz	80	1	0.5
Sweet potatoes, stage 2	4 oz	80	1	0
Vegetable beef dinner, stage 2	4 oz	80	3	2.5
Peaches, stage 2	4 oz	75	1.1	0.4
Apples and blueberries, stage 2	4 oz	70	0.2	0.2
Beef and gravy, stage 2	4 oz	70	8	2.5
Pears, stage 2	4 oz	70	0.3	0.2
Apple strawberry banana, stage 2	4 oz	60	0	0
Carrots, stage 2	4 oz	50	1	0
Applesauce, stage 2	4 oz	45	0.2	0.2
Peas, stage 2	4 oz	45	3	0
Applesauce and pineapple, stage 2	4 oz	40	0.1	0.1
Squash, stage 2	4 oz	40	1	0
Arrowroot cookie	1 cookie	22	0.4	0.7
Oatmeal	1 Tbsp (dry)	13	0.4	0.3
Rice cereal	1 Tbsp (dry)	10	0.2	0.1
Mixed grain cereal	1 Tbsp (dry)	9	0.3	0.1

Step-Down/Acute-Care Feeding

After transfer from the ICU, steps can be undertaken to convert feeding schedules from continuous to intermittent/bolus. Prior to transitioning to bolus feedings, small bowel feeding tubes should be pulled back so the distal tip is in the stomach. One successful transitional approach is to provide 3 hours' volume of feeds over increasingly shorter durations of time using a feeding pump. For example, continuous feeds of 15 mL/hr can be condensed to an intermittent feeding of 23 mL/hr for two hours and then 45 mL/hr for 1 hour. Eventually, the goal is to introduce oral feedings when the infant is clinically ready. In some cases, however, fatigue, anorexia, or swallowing dysfunction may preclude safe

or adequate feedings (12,14). Oral skill development may be delayed due to prolonged hospitalization or intubation. Infants with oral feeding difficulty should receive a consult from a feeding specialist. These therapists may employ techniques such as chin support,

Table 13.
Sample nutrition-related home monitoring
"red flag" criteria (39)

- Weight loss of 30 g in one day
- Failure to gain 20 g per day for three days
- 2 episodes of vomiting
- Loose stools (change from baseline)

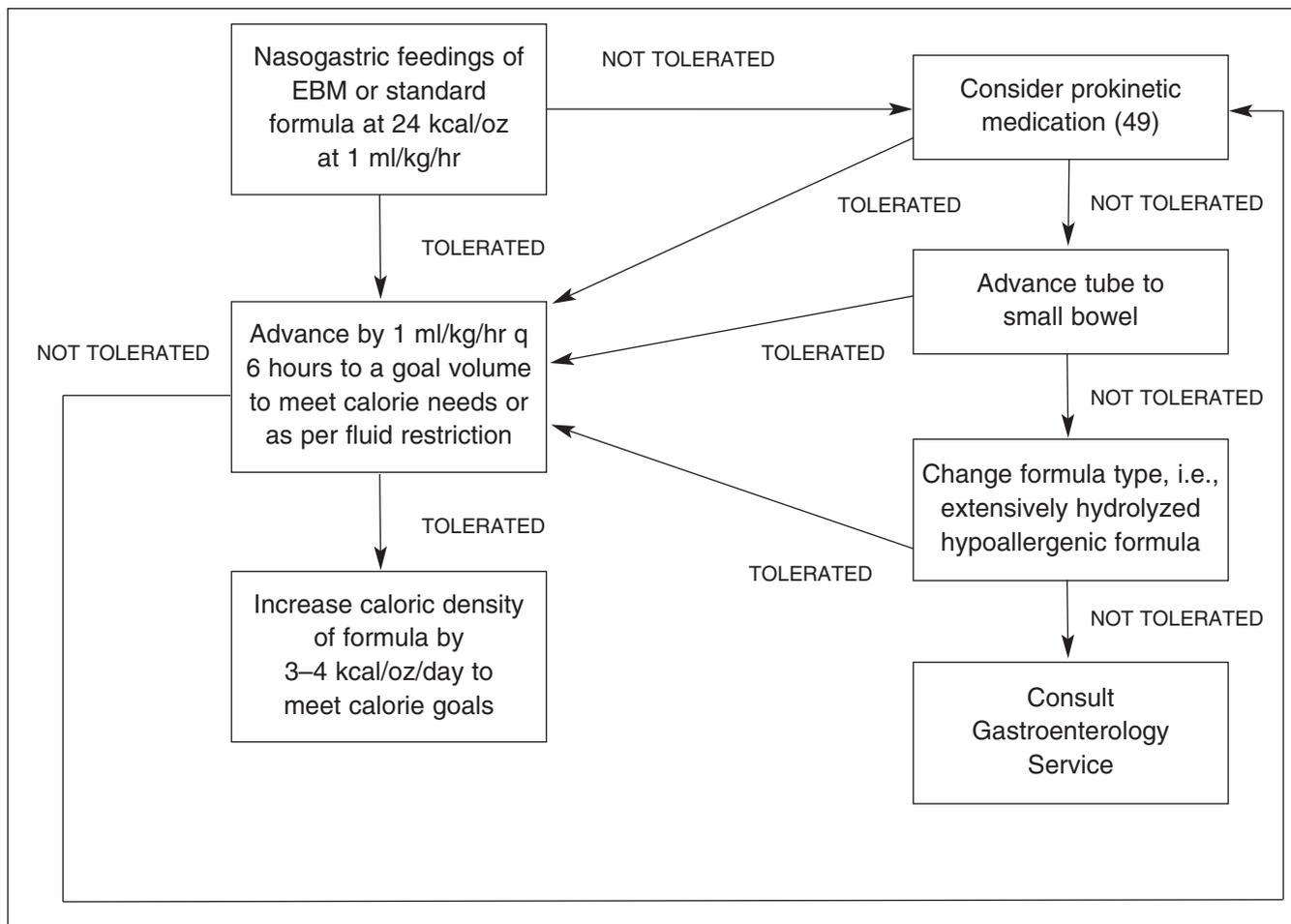


Figure 1. Guidelines for the initiation and advancement of EN for the pediatric CHD patient (49). Abbreviations: EBM = expressed breast milk

slow-flow nipples, or alternate feeding positions (such as feeding with the right side down in the case of concern for left vocal cord injury) (14). Infants who are not safe for oral feeding may benefit from practice with non-nutritive sucking from a pacifier.

After transfer from the ICU setting, some patients still require supplemental EN. For small infants in which every three hour feeds would be age-appropriate, an acceptable strategy is to allow oral trials for 20–30 minutes, then deliver the remainder of the feeding through an NGT. For older infants and children who would not be expected to feed through the night, but who are unable to take adequate nutrition during the day, nocturnal continuous drip feeds of approximately 50% estimated needs over 10–12 hours would be reasonable. Daytime bolus feeds can be added, if needed,

to supplement oral intake (14). Infants ≥ 6 months of age should be offered solid foods (preferably calorically dense foods) from a spoon, as long as their oral-motor skills are appropriate for solid food feeding. Although infants with CHD may be considered “nutritionally fragile,” as long as there is no overt contraindication, they should be encouraged to meet age-appropriate feeding milestones. Table 12 delineates the caloric content of common infant foods, which can be helpful in advising parents how to maximize nutrient content of all food intake. Older toddlers and children may benefit from a high-calorie oral supplement, such as Pediasure[®] (Abbott Nutrition, Columbus, OH) or Resource[®] Breeze (Nestle Nutrition, Florham Park, NJ), if oral intake is limited or if poor growth is a problem.

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Table 14.
Nutrition-related complications and comorbidities in the pediatric CHD patient (26,38,40,46,52–56)

<i>Complication & definition</i>	<i>Incidence</i>	<i>Etiology related to CHD</i>
Laryngeal dysfunction (vocal cord paralysis or paresis, potentially resulting in swallowing dysfunction and tracheal aspiration)	1.7%–9% after cardiac surgery Up to 50% with swallowing dysfunction	Operative trauma to the recurrent laryngeal nerve Prolonged intubation affecting vocal cords Neurological injury or prematurity affecting swallow
Acute renal failure (50% increase in serum creatinine from baseline, a decline in UOP to <0.5–1 mL/kg/hr, and/or an increase in BUN)	1%–42% after cardiac surgery	Ischemic and inflammatory insults that occur during CPB; longer CPB times are associated with higher risk Higher risk in smaller children <2 years of age, since maximal glomerular filtration rate has not yet been reached
Necrotizing enterocolitis (bowel compromise and potential necrosis in the neonatal period due to mucosal insult, injury, or ischemia worsened by bacterial proliferation)	3.3%–6.8%	Poor gut perfusion due to diastolic flow reversal in the SMA, particularly in single ventricle or left ventricular outflow tract obstruction physiology (16) Deep hypothermia during CPB causing decreased gut perfusion (16) Ischemia/reperfusion injury associated with CPB Altered intestinal barrier Proinflammatory response to CHD and CPB
Chylothorax (accumulation of chyle, which transports LCTs in the lymphatic system, in the pleural space after damage to the thoracic duct)	2.5%–4.7%	Injury to the thoracic duct during surgery Thrombosis of the superior vena cava resulting in occlusion Elevated central venous pressure following surgery

Abbreviations: UOP = urine output; BUN = blood urea nitrogen; CPB = cardiopulmonary bypass; PN = parenteral nutrition; CRRT = continuous renal replacement therapy; PD = peritoneal dialysis; SMA = superior mesenteric artery; LCT = long chain triglycerides; MCT = medium chain triglycerides; EFAD = essential fatty acid deficiency; IV = intravenous

Formula manufacturers:

- 1 – Abbott Nutrition, Columbus, OH (www.abbottnutrition.com; 1-800-227-5767)
- 2 - Vitaflo USA, Alexandria, VA (www.vitaflousa.com; 1-888-VITAFLO)
- 3 – Mead Johnson Nutritionals, Evansville, IN (www.mjn.com; 1-800-BABY-123)
- 4 – Nestle Nutrition, Florham Park, NJ (www.nestle-nutrition.com; 1-800-422-ASK2)

Nutrition-related management

Consider pre-feeding screening for those at high risk (aortic arch reconstruction)
 Consult feeding specialist
 Thicken feeds to nectar or honey consistency
 Feeding positioning (right side down)
 Supplemental/sole tube feeding

Fluid restriction (more than usual) may be required; concentrate formula caloric density or PN

Choose a formula low in electrolytes, such as Similac[®] PM 60/401 (infant), Renastart^{™2} (pediatric), or Nepro^{®1} or Suplena^{®1} (adult)

If no dialysis:

If on, PN reduce trace element delivery by 50%
 Provide 1.5 g/kg/day protein

If dialysis is required:

Supplement with water soluble vitamins and trace elements, including selenium
 Provide 2–3 g/kg/day protein for CRRT and 4 g/kg/day for PD

Provision of adequate PN if feeds are held
 When feeds are restarted, use EBM or hypoallergenic infant formula at 20 kcal/oz; begin and advance conservatively

Limit dietary LCT, often for up to 6 weeks after surgery: provide very high MCT formula (i.e., Enfaport^{®3}) for infants or a diet very low (<10 g/day) in total fat for toddlers and children; tube-fed children can receive Enfaport^{®3}, Portagen^{®3}, or Tolorex^{®4}

Breast milk can be skimmed with a centrifuge, then fortified with high-MCT formula, carbohydrate modules, or MCT oil
 Ensure 2–4% of calories from long chain fat to prevent EFAD (provide controlled doses of LCT-containing oil or IV lipid)
 Ensure adequate protein, electrolyte, and vitamin intake, as nutrients are lost in chyle drained via chest tube
 PN (including IV lipid) may be required in cases refractory to nutrition and medical management

Current literature suggests that feeding at the breast can be an option for CHD infants, although traditionally breastfeeding has been thought to burn more calories than bottle feeding. However, research shows that severity of CHD is not a predictor of breastfeeding success (36). Furthermore, oxygen saturations can be maintained and can be more consistent in an infant feeding at the breast than the same infant feeding from a bottle, indicating lower cardiorespiratory stress when feeding at the breast (37). Although it is not as easy to measure actual intake with breastfeeding, measuring weights before and after nursing sessions using an electronic scale can be accurate in measuring intake (38). A lactation consultant is a valuable addition to the care team when an infant is feeding at the breast or when a mother is pumping EBM.

Discharge to Home

Prior to discharge from the hospital, a patient's feeding schedule should be maximized to promote nutrition intake and patient as well as family quality of life. Caregivers should demonstrate competency in mixing infant formula to the required caloric density and should be able to verbalize appropriate storage and handling techniques to reduce risk of foodborne illness. Caregivers must also demonstrate understanding of feeding schedule and delivery methods and of how to monitor for and troubleshoot complications. The clinician should ensure the family has a reliable way to obtain formula; in many circumstances, formula can be obtained from the Special Supplemental Nutrition Program for Women, Infants, Children (WIC program). However, WIC may not be able to provide 100% of a child's required formula. If a family is not eligible for the WIC program or if the WIC program cannot provide enough formula, health insurance may cover costs if the patient requires tube feeding.

For infants at high nutrition and medical risk, such as those with HLHS in between their first and second surgeries, home surveillance programs may be beneficial in monitoring nutrition progress. In such programs, families are given strict instructions on "red flags" (see Table 13) that would necessitate contact with the medical team. Families may be asked to monitor oxygen saturations, weights, and formula adequacy on a daily

basis at home and then report these results on a regular basis to a member of the medical team. Although these programs increase burden on families, they increase interstage survival and reduce malnutrition (39).

Nutrition Management of Complications and Comorbidities

Unfortunately, many nutrition-related complications and comorbidities accompany CHD and cardiac surgery. For instance, the incidence of NEC has been reported to be 10–100 times higher in infants with CHD than in those without. NEC is commonly thought to be a disease of prematurity, but up to 10% of cases occur in term infants (40). Emerging evidence indicates that NEC in the CHD infant is of a different etiology than in the premature infant, although the clinical sequelae are similar. Acute renal failure, laryngeal and swallow dysfunction, and chylothorax are additional complications related to nutrition. Table 14 summarizes etiologies of these complications as they relate to the CHD infant and how these comorbidities may be managed nutritionally.

SUMMARY

In most cases, CHD prove to be a challenge for the clinician because of the complex interplay between medical, surgical, nutritional, and social factors. 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. As such, many infants require supplemental EN or PN, although the ultimate goal is commencement or resumption of oral feeding. Other factors, such as NEC, chylothorax, and laryngeal and neurological dysfunction, play a major role in the requisite nutrition therapy for the CHD infant. 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. ■

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