NUTRITION OUTCOMES IN INFANTS AFTER SURGERY FOR CONGENITAL HEART DEFECTS

Brandis A. Roman

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Thesis Committee:

Leslie Hildebrandt, Ph.D.	Committee Chair
Roschelle Heuberger, Ph.D.	Faculty Member
John Logomarsino, Ph.D.	Faculty Member
May 30, 2012	Date of Defense
Roger Coles, Ed.D.	Dean College of Graduate Studies
July 12, 2012	Approved by the College of Graduate Studies

This project is dedicated to my
pediatric congenital heart defect patients
and their families,
who have taught me about
the true meaning of joy, resilience, and love.

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ABSTRACT

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by Brandis A. Roman

Congenital heart defects (CHD) are the most common birth defect in the United States, affecting 1% of all pregnancies. Historically, efforts have focused on reducing mortality during surgical palliation or correction, but, with the advent of modern surgical techniques, mortality has improved, and thus the focus has shifted to improvement in nutrition status and quality of life. Infants with CHD are at nutrition risk for a variety of factors, including growth retardation en utero, poor caloric intake, increased energy expenditure, and gastrointestinal morbidity. The postoperative period after CHD surgery is a time of high nutritional morbidity, with limited nutrition intake and high energy needs, which can affect length of stay and hospital costs. The purpose of this project was to examine postoperative nutrition outcomes in 100 infants less than one year of age during 130 hospitalizations for CHD surgery to determine potential areas for improvement in nutrition-related management. Subjects were selected randomly from an institutional database, and a retrospective chart review was undertaken. Mean weight-for-age z-score significantly worsened from birth to preoperative admission and from admission to hospital discharge for all study infants and for those specifically with hypoplastic left heart syndrome, a diagnosis with high morbidity and mortality. In multivariate analysis, worsening of weight-for-age z-score was not related to intensive care unit or total hospital length of stay. The incidence of necrotizing enterocolitis, a complication with significant consequences related to hospital stay, cost of care, nutrition status, and mortality, was 10% in the entire study group. Other studies have found value, including a reduction in necrotizing enterocolitis incidence, with the

development of a standardized approach to feeding the postoperative CHD infant using a feeding protocol or algorithm. This could be a viable strategy to reduce variation and improve outcomes at the author's institution.

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CHAPTER I

INTRODUCTION

Congenital heart defects (CHD) are structural aberrations in the heart and/or great vessels, which result in altered cardiovascular function; these defects are a result of abnormal intrauterine development (1, 2). CHD are the most common and deadly birth defect in the United States, with a prevalence of 0.9-1.4% (1, 3-5). Appendix A describes some common defects.

Many infants and children require surgical palliation or correction of their defect, oftentimes in the first year of life, in order to restore pulmonary and systemic circulations to improve chances of survival, quality of life, and growth (1). Surgical techniques and medical management have reduced infant CHD-related mortality since the first pediatric open-heart surgery was done in 1944. As such, the focus of much research and attention is on preserving and improving the nutrition status of these high-risk children in the perioperative period (6). Unfortunately, the postoperative period after CHD surgery is a time of particularly high nutritional risk due to many factors, including preoperative (baseline) malnutrition, barriers to postoperative nutrition in the intensive care unit (ICU), and increased energy and nutrient needs postoperatively. Poor nutrition in the cardiac surgery patient can have multiple deleterious consequences, including prolonged length of stay (LOS) and increased hospital costs.

Many infants and children enter the operating room already malnourished (7) because of factors related to the heart defect. Multiple studies have observed that mean weights (8-11) and heights (11) of infants with uncorrected CHD are significantly lower than age-matched controls. Authors report overall prevalence of malnutrition of up to 90% using height and/or weight parameters in mixed-defect cohort and cross-sectional studies (12-31). Measured decrease in fat and muscle mass (32-34) has been reported in up to ~20% of children with CHD (16, 20), and a

reduction in visceral organ size (brain, thymus, adrenal glands, liver, and kidney) compared to controls has been noted upon autopsy of infants with CHD (33). Estimates may not adequately reflect the true prevalence of malnutrition in the CHD population, as most studies exclude infants who were premature or small for gestational age, those who have comorbidities, such as genetic defects, or those undergoing palliative procedures. However, these numbers may also overestimate the true prevalence of malnutrition, as most infants studied are those who are hospitalized or being considered for heart surgery, which are those with the most significant lesions, which likely impact nutrition status more than milder defects.

The etiology of malnutrition in pediatric CHD patients is complex and multifactorial. In many cases, growth failure is probably due to a combination of inadequate intake, inappropriate assimilation and/or absorption of nutrients, and increased energy expenditure (35, 36). Additionally, there is some evidence that infants with CHD have intrauterine growth retardation and are born at a birth weight lower than the norm. Genetic syndromes, of which CHD are known to be a part, can also place a child at increased risk for growth and development problems.

Regardless of etiology, malnutrition can contribute to increased morbidity, mortality, and hospital costs in the postoperative period. Baseline malnutrition has been associated with an increase in hospital LOS in infants and children with CHD (37, 38), while higher preoperative weights have been independently correlated with shorter ICU LOS (39). In a report of 310 children undergoing even low surgical risk CHD procedures, failure to thrive increased hospital charges by $13,151 per patient ($p=0.02$, compared to those without failure to thrive) (38). Malnutrition has been associated with an increased length of mechanical ventilation (40) and increased quantity of care requirement (41) in the pediatric ICU population. Lower preoperative

weight has also been associated with an increased risk of mortality during the perioperative period in CHD infants (42, 43).

In the postoperative period, many challenges exist to optimizing nutrition status, including increased energy needs after surgery and/or to compensate for baseline malnutrition. Inadequate intake due to fluid restriction, multiple medical and airway procedures, difficulty with oral intake, and gastrointestinal complications make meeting those higher energy needs sometimes impossible. Poor weight gain, malnutrition, and prolonged attainment of full enteral feeds postoperatively can contribute to increased postoperative (44) and ICU (45, 46) LOS and mechanical ventilation requirements (40), which can all directly affect health care costs.

The purpose of this retrospective study was to describe the postoperative, hospital-based nutrition outcomes of a heterogeneous group of CHD infants at a tertiary care medical center/teaching hospital with pediatric cardiothoracic surgery and cardiology programs. These data were compared with similar outcome measures from other pediatric cardiology programs to identify areas for improvement in current postoperative nutrition practices.

CHAPTER II

METHODS

Subject Selection

Potential subjects were identified from an institutional clinical data repository of patient health information and outcomes using search parameters corresponding to the following inclusion criteria: 1) International Classification of Disease version 9 (ICD-9) codes corresponding to CHD; 2) patients ≤12 months of age; 3) those with an inpatient stay and a cardiothoracic procedure between the dates of January 1, 2005 and December 31, 2010. Subjects were excluded if they had an isolated non-structural cardiac abnormality (such as dysrhythmias), did not undergo a palliative or corrective procedure for CHD or did so after one year of age, or died prior to the commencement of enteral feedings. For the purposes of this study, a patent ductus arteriosus (PDA) was considered transitional fetal physiology and was not considered to be a defect; however, infants with a PDA were included in the study if they had a co-existing defect of hemodynamic significance, including those defects presented in Appendix A. Infants undergoing interventional cardiac catheterization for CHD were included as long as they were admitted to the ICU post-procedure and had a post-procedure LOS of at least 48 hours.

The repository search yielded 1728 potential matches. Three hundred patients were excluded because they did not fulfill inclusion criteria. Out of the remaining 1428 patients, 100 subjects were randomly chosen for the study using a random numbers table.

Data Collection and Analysis

After approval from the Institutional Review Boards of both Central Michigan University and the University of Virginia, both of which granted a waiver of consent for data collection, the

electronic medical record of each patient was accessed to abstract relevant data. Discharge summaries, operative notes, progress notes, and bedside flow sheets were reviewed for each patient. Data were collected regarding birth (weight, length, APGAR scores at one and five minutes, estimated gestational age, type of cardiac lesion, and the presence of genetic syndromes), preoperative admission (age, weight, length, preoperative feeding, and need for prostaglandin infusion), surgical procedure (age, weight and length at which the procedure was done, type of procedure, and need for cardiopulmonary bypass [CPB]), postoperative ICU admission (ICU LOS, days to first enteral or oral feeding, type and method of enteral or oral feeding, complications, PRISM score upon ICU admission, highest whole blood lactate value during ICU stay, lowest pH value on arterial blood gas measurement during ICU stay, need for and duration of parenteral nutrition, and duration of mechanical ventilation), and discharge status (weight and length, total hospital LOS, day at which full enteral feeds was attained, feeding at discharge, and need for feeding tube). The APGAR score assesses Appearance, Pulse, Grimace, Activity, and Respirations at one and five minutes after birth and has been used quite commonly since the 1950s to quickly assess the condition of a newborn in the immediate post-delivery period (47). The PRISM score is the Pediatric Risk of Mortality III score, which has been validated for use in the pediatric ICU setting. This score takes into account physiological variables, such as cardiovascular vital signs, blood gas results, blood chemistry, and hematological parameters, to objectively quantify level of critical illness and risk of death (48). In this study, the score was calculated at the time of admission to the ICU from the operating room. Whole blood lactate and arterial blood pH were measured as surrogate markers of cardiac output and tissue oxygen debt. Full enteral feeding was defined as a volume required to provide 100 kcal/kg/day (volume varied depending on caloric density of feeding fluid). While this is

likely less than what most patients would require for adequate growth or catch-up growth, this is a well-accepted calorie requirement for most infants during the first year of life, and, given the heterogeneity of this sample, was deemed a reasonable standard to meet at least 75-80% of the calorie needs of most patients. If an infant required more than one hospitalization for a cardiac procedure during the first year of life, information was collected for each hospitalization. Weight and length were accepted as a preoperative values if they were measured at the time of hospital admission or during a preoperative clinic visit the day before admission. Day of surgery weight and length were accepted as such if the measurements occurred the morning before surgery or during a preoperative clinic visit the day before the surgery. If weight and length were not measured on the day of discharge, a weight on the day before discharge was accepted and a length within the past 7 days was accepted. Duration of ventilation was rounded to the nearest half hour, and parenteral nutrition duration was rounded to the nearest hour.

Weight-for-age (WFA), length-for-age (LFA), and weight-for-length (WFL) z-scores for infants with estimated gestational age $\geq$ 37 weeks were determined using EpiInfo version 7.0.9.0 (Centers for Disease Control, Atlanta, GA). This program utilizes the World Health Organization growth standards to determine anthropometric z-scores. WFA and LFA z-scores for infants with estimated gestational age <37 weeks were determined using data from the Fenton growth curves (49) until the infants were 50 weeks post menstrual age. WFL z-scores were not available for these infants. After 50 weeks post menstrual age, z-scores were determined using EpiInfo, adjusting manually for prematurity.

Data were analyzed using the Statistical Package for the Social Sciences version 20 (SPSS, Chicago, IL). Normality of continuous data was determined using the Shapiro-Wilk test. Measures of central tendency for normally distributed data were expressed as mean $\pm$ standard

deviation, while the median and range were used for non-normally distributed data. To assess the change in anthropometric z-scores from birth to hospital admission and from hospital admission to discharge, paired samples t-test was used to compare z-scores at various time points for normally distributed z-scores. Non-normally distributed z-scores at various time points were compared using the Wilcoxon signed rank test. To investigate how deterioration in nutrition status (change in anthropometric z-scores) could affect LOS, Kendall's tau correlation coefficient was used to measure correlations between change in anthropometric z-scores during hospitalization and ICU and total hospital LOS. Statistical significance was set at the 0.05 level.

Because of the heterogeneity of the sample with regard to specific lesion and surgical procedure, extensive subgroup analysis was not possible. However, because of available comparison groups in the current literature and perceived nutritional fragility of these patients (50-53), a subgroup analysis was undertaken for infants undergoing stage I palliation for hypoplastic left heart syndrome (HLHS).

CHAPTER III

RESULTS

All Infants

Birth characteristics of study subjects are reviewed in Table 1. The majority of infants were born at term gestation ($\geq$ 37 weeks), although 23% were born at 36 weeks gestational age or less. The median gestational age was 38 weeks; 18 infants were recorded as being at "term" gestation without a specific number of weeks available in the medical record. The most common heart lesion was HLHS (21%), followed by Tetralogy of Fallot (15%), coarctation of the aorta (11%), and septal defects of the ventricle and/or atria (11%). Refer to Appendix A for an explanation of the physiology of each defect. Twenty infants were also diagnosed with a chromosomal abnormality, the most common (11%) being trisomy 21. Table 2 describes birth anthropometrics. Mean WFA z-score was -0.31 $\pm$ 1.13, corresponding to a percentile between 0 and the 25th. Mean LFA z-score approached zero (-0.08 $\pm$ 1.41), whereas mean WFL z-score was closer to the 25th percentile at -0.52 $\pm$ 1.31.

Data were available on 130 hospital admissions. Twenty-eight patients presented for two hospitalizations involving procedures for palliation or repair of a CHD during the first year of life, whereas one patient presented for three admissions. The median age at admission was 56 days, although 58 (44.62%) admissions occurred in the neonatal period (<30 days of life). Mean WFA z-score at admission for a procedure was -0.79 $\pm$ 1.22, which was significantly less than birth WFA z-scores (p=0.000). Mean LFA and WFL z-scores at admission were -0.49 $\pm$ 1.53 and -0.67 $\pm$ 1.21, which were not significantly different from birth z-scores (Table 2). Most infants (44.6%) were fed orally immediately preceding their cardiac procedures, although approximately one quarter received parenteral nutrition only.

Table 1. Birth Characteristics of Subjects

Estimated gestational age (weeks), median, range (total n)	38, 13 (81)*
APGAR score at one minute, median, range (total n)	8, 8 (60)
APGAR score at five minutes, median, range (total n)	9, 4 (60)
Gender, male, % (total n)	62% (100)
Parity, %, (total n)	(100)
Singleton	90%
Twin	9%
Triplet	1%
Type of defect, %, (total n)	(100)
Hypoplastic left heart syndrome	21%
Tetralogy of Fallot	15%
Coarctation of the aorta	11%
Septal defects	11%
Atrioventricular canal	9%
Transposition of the great arteries	8%
Double outlet right ventricle	8%
Other complex single ventricle	6%
Total or partial anomalous pulmonary venous return	5%
Valve stenosis	4%
Aortopulmonary window	1%
Ebstein's anomaly	1%
Chromosomal abnormalities, %, (total n)	(100)
Trisomy 21	11%
22q11 deletion	3%
Turner syndrome	2%
Other	4%

APGAR = Appearance, Pulse, Grimace, Activity, Respirations
*18 infants were recorded as being born at "term gestation" without an estimated gestational age in weeks

At the time of surgery, at a median of 58 days of age, the mean WFA, LFA, and WFL z-scores were similar to those at hospital admission (-0.80, -0.52, -0.63, respectively; Table 2). Approximately 60% of hospitalizations involved a palliative procedure, whereas the other 40% involved complete correction of the defect (Table 3). The most common palliative procedures were the bidirectional Glenn procedure, the Norwood procedure, and Blalock-Taussig shunt placement, whereas the most common complete surgical corrections were closures of atrial and/or ventricular septal defects, excision of coarctation of the aorta, and complete repair of

Tetralogy of Fallot. Most procedures (70%) required CPB. Appendix B explains each surgical procedure.

Table 2. Age and Anthropometric Measurements of Subjects*

	Birth	Pre-operative admission	Surgery	Discharge, transfer, or death
Age (days)	-	56.27, 343 (130)	58.30, 343 (130)	79.5, 340 (130)
Weight (kg)	3.12, 3.41 (89)	4.07, 8.09 (130)	4.06, 8.09 (129)	4.10, 8.04 (125)
Weight-for-age z-score	-0.31 ± 1.13 (89)	-0.79 ± 1.22[a] (130)	-0.80 ± 1.19 (129)	-1.07 ± 1.23[b] (125)
Length (cm)	49, 21 (58)	53.5, 38.8 (121)	54, 38.8 (119)	55, 31 (105)
Length-for-age z-score	-0.08 ± 1.41 (58)	-0.49 ± 1.53 (121)	-0.52 ± 1.49 (119)	-0.70, 8.73 (105)
Weight-for-length z-score	-0.52 ± 1.31 (44)	-0.67 ± 1.31 (101)	-0.63 ± 1.34 (100)	-0.61, 7.41 (91)

* Values given as mean ± SD (total n) when normally distributed or as median, range (total n) when non-normally distributed
a Significantly different from birth weight-for-age z-score, $p<0.01$
b Significantly different from weight-for-age z-score at admission preoperatively, $p<0.01$

In the postoperative period, 50.8% of admissions necessitated parenteral nutrition, with a median duration of 109 hours (range: 2545 hours, n=66). Tube and/or oral feeds were started on median postoperative day 2.03 (range: 16 days), and goal tube and/or oral feeds were reached a median of four days later (range: 149 days, n=111). Almost half (49.2%) of patients were able to take oral feeds as part of their first feedings, although 77 patients (59.2%) required tube feedings (either via hospital-placed nasogastric or nasoenteric tubes or previously placed gastrostomy tubes) during the hospital stay for sole or supplemental nutrition. Of note, three infants in the postoperative period were trialed on oral *ad libitum* feedings, but eventually required nasogastric tube placement for continuous feedings due to inadequate volume intake or other clinical factors.

Table 3. Surgical Characteristics of Infants Undergoing Palliation or Surgical Correction

Palliative procedures, % of total	59.23%
Bi-directional Glenn procedure	18.46%
Norwood with Sano modification	13.85%
Blalock-Taussig (BT) shunt	12.31%
Damus-Kaye-Stansel procedure	4.62%
Other	3.08%
Central shunt	1.54%
Hybrid procedure	1.54%
Norwood with modified Blalock-Taussig (BT) shunt	1.54%
Rastelli procedure	1.54%
Corrective procedures, % of total	40.77%
Ventricular and/or atrial septal defect repair	9.23%
Coarctation of the aorta repair	7.69%
Tetralogy of Fallot repair	7.69%
Arterial switch	4.62%
Atrioventricular canal repair	4.62%
Total or partial anomalous venous return repair	3.85%
Other	3.85%
Required cardiopulmonary bypass, %	70%

As shown in Table 4, standard cow's milk protein formula was used most often (51.5%), although approximately one third of infants received some form of breast milk. Most infants were started on a standard caloric density formula (20 kcal/oz), although 21.5% received 24 kcal/oz feedings as their first tube and/or oral feeding. Continuous feedings were the most common initial delivery method at first feeding (48.5%). Feeds were held for a median of 8 hours (range: 2611 hours) per patient at any point during the postoperative stay. One patient had his enteral feeds held for greater than 2000 hours, and 16 patients had their feeds held for 100 or more hours.

Out of those surviving the hospitalization, 117 were discharged to home, and 8 were transferred to an outside hospital (Figure 1). Most patients (86.7%) were able to achieve goal enteral feedings ($\geq$ 100 kcal/kg/day) prior to discharge, transfer to outside hospital, or death. Out

Table 4. Nutrition Delivery in Infants Undergoing Palliation or Surgical Correction at First Feeding Postoperatively and at Discharge, Transfer, or Death

	First feeding postoperatively	Discharge, transfer, or death
Feeding fluid used, % (total n)	(130)	(126)
Standard cow's milk based	51.5%	38.1%
Expressed breast milk or breast feeding	36.2%	24.6%
Extensively hydrolyzed protein	3.8%	4.8%
Premature transitional	2.3%	4.0%
Combination of expressed breast milk/breast feeding and formula	1.5%	10.3%
Soy protein based	1.5%	2.4%
Elemental	0.8%	0%
Fat modified	0.8%	7.1%
Lactose free cow's milk based	0.8%	2.4%
Premature	0.8%	3.2%
Added rice	0%	2.4%
Toddler	0%	0.8%
Caloric density (kilocalories per ounce), % (total n)	(130)	(126)
20	72.3%	33.3%
24	21.5%	44.4%
22	2.3%	1.6%
27	2.3%	17.5%
26	0.8%	0.8%
30	0.8%	2.4%
Feeding schedule	(130)	-
Continuous	48.5%	-
Oral *ad libitum**	43.1%	-
Bolus/intermittent	7.7%	-
Combination bolus/intermittent (daytime) and continuous (nocturnal)	0.8%	-

* In three instances, patients were offered oral *ad libitum* feedings, but were subsequently transition to continuous nasogastric feedings.

of the 71 patients who required nasogastric or nasoenteric feedings during their post-procedure stays, more than two-thirds (68.5%) were able to have their temporary tubes removed (Figure 2), which occurred on median postoperative day 10.14 (range: 35 days, n=48). Three patients required placement of gastrostomy tubes for persistent feeding problems.

Figure 3 depicts incidence of complications postoperatively. The median ICU and total LOS were 5.07 (range: 75 days, n=112) and 13.18 days (range: 172 days, n=130), respectively. Correlates of ICU and hospital LOS are given in Table 5. PRISM score upon ICU admission postoperatively (median: 6, range: 17, n=129), highest lactate during ICU stay (median: 3.11, range: 17.63, n=127), and total duration of ventilation (median: 48.5 hours, range 1389 hours, n=129) were all significantly positively correlated with ICU and hospital LOS. In contrast, lowest pH during ICU stay (median: 7.292, range: 0.632, n=129) was negatively correlated with ICU and hospital LOS. Changes in WFA, LFA, and WFL z-scores were not found to have significant associations with LOS, although change in WFA z-score trended towards significance in an inverse relationship to total hospital LOS (p=0.079).

At discharge to home, death, or transfer to an outside hospital, the mean WFA z-score was -1.07 (Table 2), which was significantly different from that at hospital admission (p=0.000). LFA and WFL z-scores were not significantly different from preoperative admission values. The median weight change during the hospitalization was net positive 60 grams (range: 385 grams, n=125), although one third of patients (33.8%) lost weight during their stay. Out of 117 patients discharged to home, nine (7.7%) required supplemental or full nasogastric tube feedings, six (5.1%) were fed with a previously placed gastrostomy tube, and three (2.61%) were using a newly placed gastrostomy tube (Figure 2). The most common feeding fluid at discharge was standard cow's milk based formula, although ~35% of infants were receiving at least some part of their nutrition as breast milk. Most infants were discharged on a calorically dense formula or fortified breast milk (Table 4).

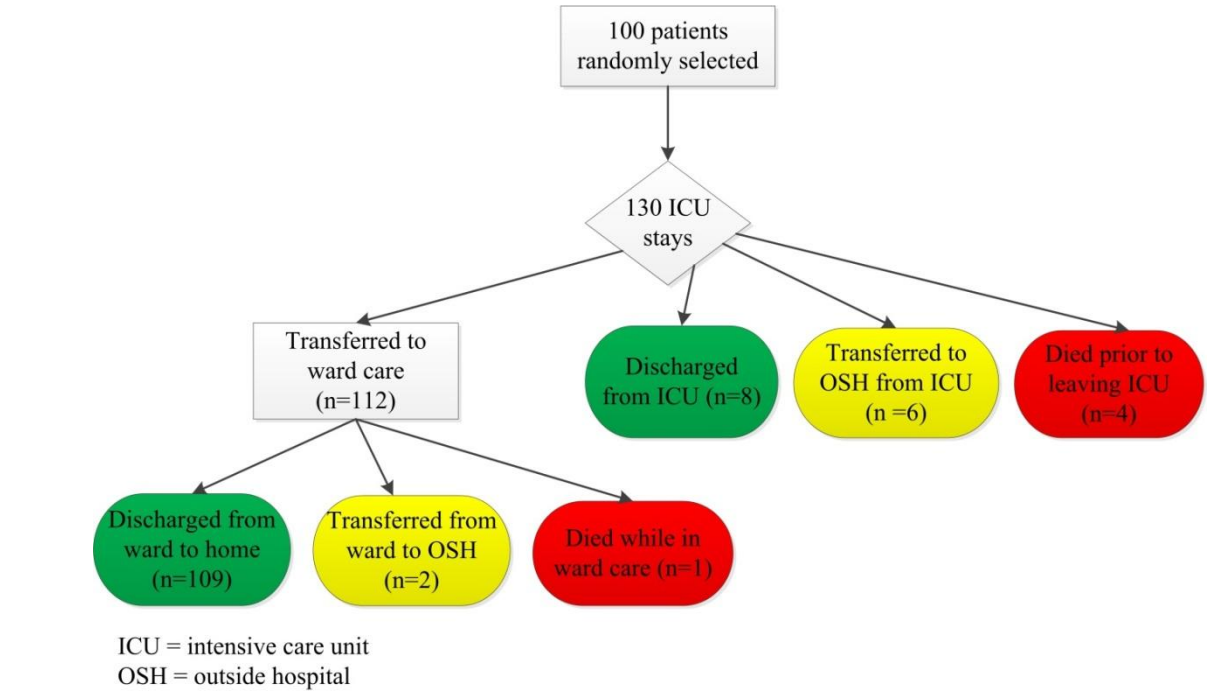


Figure 1. Subject Progression From Selection to Discharge, Transfer, or Death

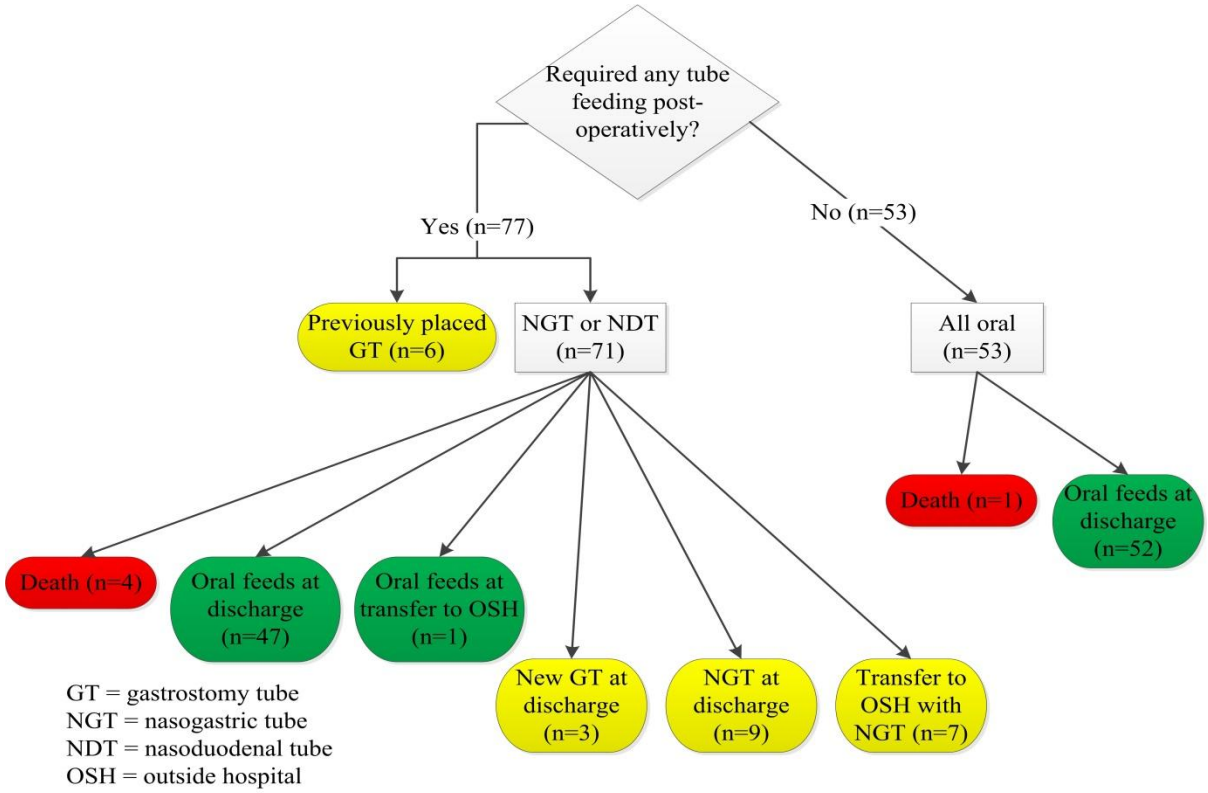


Figure 2. Feeding Modality Postoperatively and at Time of Discharge, Transfer, or Death

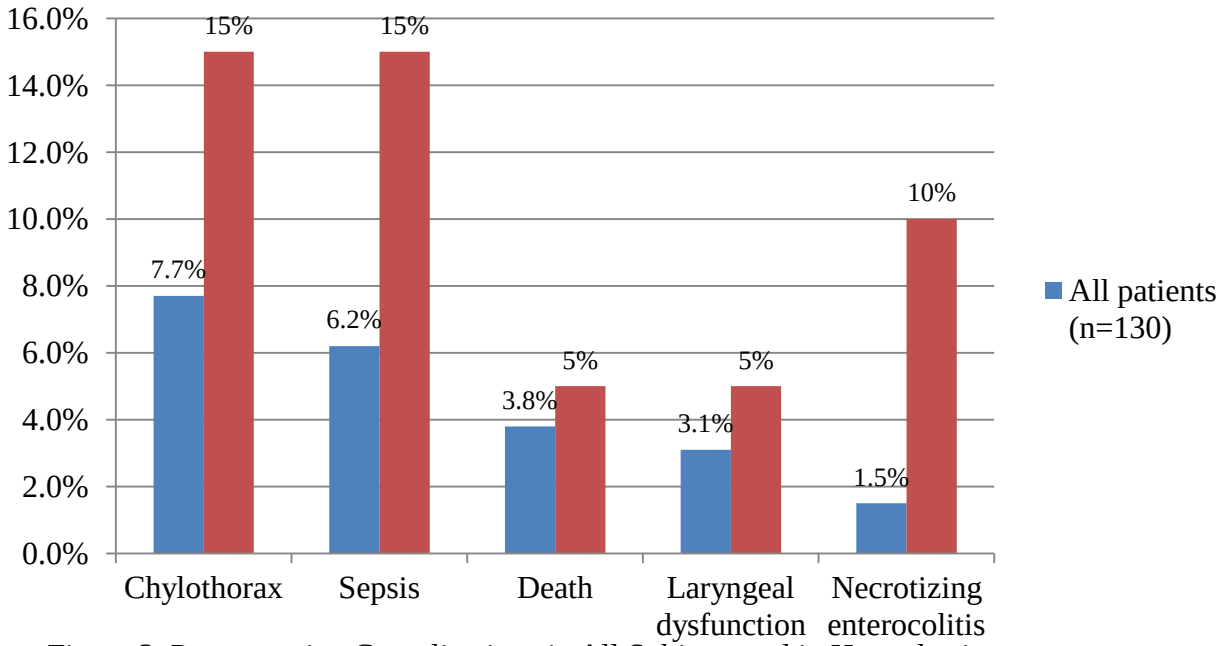


Figure 3. Postoperative Complications in All Subjects and in Hypoplastic Left Heart Syndrome (HLHS) Patients

Table 5. Correlates of Intensive Care Unit and Total Hospital Length of Stay in All Subjects

	ICU LOS	Total LOS
PRISM score upon admission to ICU	0.260*	0.327*
Lowest pH during ICU stay	-0.151**	-0.169*
Highest lactate level during ICU stay	0.292*	0.281*
Duration of ventilation during ICU stay	0.631*	0.543*
Change in weight-for-age z-score during hospitalization	NS	NS
Change in length-for-age z-score during hospitalization	NS	NS
Change in weight-for-length z-score during hospitalization	NS	NS

* Significant at the 0.01 level using Kendall's tau
** Significant at the 0.05 level using Kendall's tau
LOS = length of stay
PRISM = Pediatric Risk of Mortality score
ICU = intensive care unit
NS = not significant

Infants with Hypoplastic Left Heart Syndrome Status Post Stage I Palliation

Anthropometric values and corresponding z-scores for HLHS infants undergoing stage I palliation are presented in Table 6. Infants were admitted at a median corresponding to the day of birth, with a WFA z-score of -0.46. The estimated gestational age was 38.5 weeks (range: 7 weeks, n=20). Median ICU and total LOS were 12 days (range: 72, n=19) and 23.32 days (range: 85, n=20), respectively. The median age at discharge, transfer, or death was 23 days, and the WFA z-score was significantly lower ($p<0.05$) than that at hospital admission. Median total weight change during the hospitalization was 145 grams (range: 2077, n=19) (representing an increase) and was 267 $\pm$ 587 grams on average since surgery (n=19). Average daily weight change during the hospitalization was 7.2 $\pm$ 8.49 g/day (n=19), and median daily increase since surgery was 10.17 g/day (range: 92.13, n=19).

The majority (90%) of these infants underwent the Norwood procedure with Sano modification; the other 10% underwent a hybrid procedure. All those undergoing the Norwood did so with CPB. Postoperatively, three patients developed chylothorax, three sepsis, two necrotizing enterocolitis (NEC), and one laryngeal dysfunction (Figure 3). One patient died

Table 6. Age and Anthropometric Measurements of Hypoplastic Left Heart Syndrome Subjects Undergoing Stage I Palliation*

	Preoperative admission	Surgery	Discharge, transfer, or death
Age (days)	0, 2 (20)	5.12 $\pm$ 3.11 (20)	23, 86 (20)
Weight (kg)	3.01 $\pm$ 0.52 (20)	3.12 $\pm$ 0.55 (20)	3.44, 1.45 (19)
Weight-for-age z-score	-0.46 $\pm$ 0.96 20)	-0.26 $\pm$ 0.96 (20)	-0.85 $\pm$ 1.25[a] (19)
Length (cm)	49, 13 (20)	49, 14 (19)	51.24 $\pm$ 2.47 (17)
Length-for-age z-score	-0.23 $\pm$ 1.08 (20)	-0.47, 4.3 (19)	-0.47 $\pm$ 1.67 (17)
Weight-for-length z-score	-0.51 $\pm$ 1.13 (17)	-0.16 $\pm$ 1.31 (17)	-0.64 $\pm$ 1.31 (14)

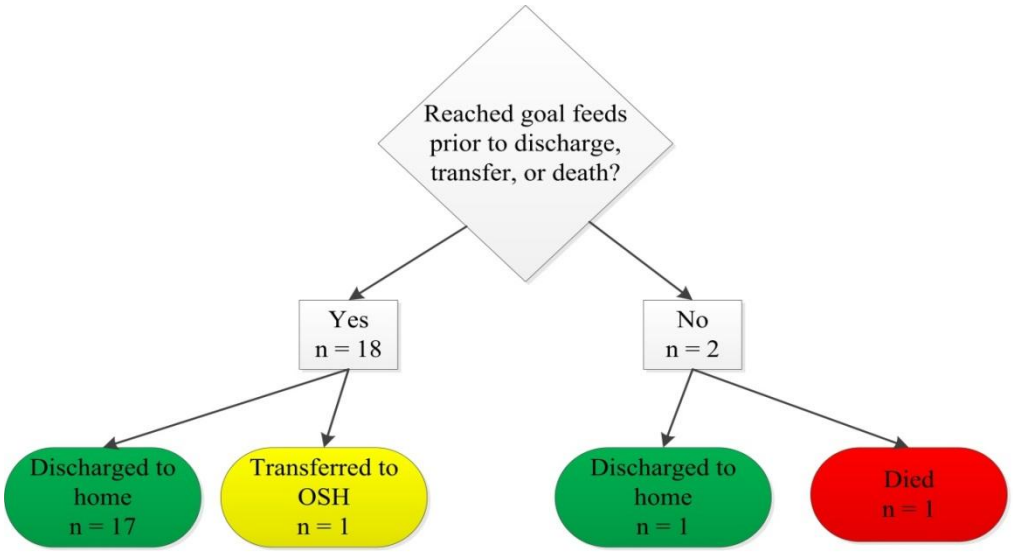
* Values given as mean $\pm$ SD (total n) when normally distributed or as median, range (total n) when non-normally distributed

a Significantly different from weight-for-age z-score at admission preoperatively, $p<0.05$

postoperatively.

All 20 patients required parenteral nutrition postoperatively, for a median of 176.5 hours (range: 1599, n=20). On average, patients were fed on postoperative day 4.2 $\pm$ 2.09 (n=20). Half of patients were fed with breast milk and the other half were fed with standard cow's milk formula. Most (80%) received standard caloric density formula (20 kcal/oz). Eighteen patients (90%) reached full enteral feeds prior to discharge to home, transfer to outside facility, or death; one patient died and one patient was discharged before achieving full enteral feeds (Figure 4). For those 18 who did reach full enteral feeds, they did so on median day 7.5 (range: 46) after beginning enteral feeds. One patient who reached full feeds was transferred to an outside hospital rather than discharged to home. Eighteen patients were discharged to home (17 who reached goal feeds and one who did not); most did not require tube feeds at home, but three (16.7%) required supplemental nasogastric feeds at discharge and two (11.1%) required placement of a gastrostomy tube prior to discharge. Patients who did not require a feeding tube at discharge had their tubes removed at postoperative day 14 (range: 28, n=13). At discharge, transfer, or death, breast milk and standard cow's milk formula remained the most common feeding fluids, but the most common caloric density was 24 kcal/oz, compared to 20 kcal/oz in the immediate postoperative period. Five patients (26.3%) required 27 kcal/oz feedings.

Change in WFA z-score from admission to discharge was negatively correlated ($r = -0.364$, $p<0.05$) with total LOS, however, this correlation did not persist after adjustment using linear regression to account for potentially confounding variables (duration of ventilation, lowest pH, and highest lactate level). There were no significant differences in admission and discharge z-scores for LFA or WFL, nor any correlation between change in LFA or WFL z-scores and ICU or total LOS.



OSH = outside hospital

Figure 4. Progression of Patients with Hypoplastic Left Heart Syndrome After Stage I Palliation

CHAPTER IV

DISCUSSION

This retrospective study of 100 CHD patients describes some of the common nutrition outcomes and challenges in the period post cardiac surgery. Postoperative malnutrition can be the result of a complex interplay between preoperative malnutrition, poor nutrition delivery in the immediate postoperative period, and high energy needs after surgery. It is difficult to know which factor is the main culprit, but it is certainly key to consider each element in these high-risk patients.

Indeed, the infants in this study exhibited an element of preoperative malnutrition. The mean WFA z-score was -0.31 at birth, representing some growth retardation in utero, which is similar to what has been presented in other studies (54-58). Despite weighing slightly less than expected for gestational age, most infants in this sample were born at term gestation, which follows what other authors have found (54, 59). It is unclear how suboptimal fetal growth and altered cardiogenesis en utero are related. It is conceivable that altered growth can affect formation of the heart, but it is also possible that altered blood flow and hemodynamics caused by CHD can affect subsequent prenatal growth and development (55). Maternal exposure to teratogens or toxins could also affect both cardiogenesis and overall growth and development (60, 61). Regardless of the connection, these infants have not only the defect to overcome to achieve optimal nutrition status, but also the lower than expected birth weight.

At the time of presentation for palliation or surgical correction, mean WFA z-score had significantly worsened to -0.79, indicating further progression of baseline preoperative malnutrition in the time period between birth and cardiac surgery. This study did not attempt to examine reasons for the deterioration of nutrition status between birth and surgical intervention;

however, one could postulate this is likely related to a variety of factors, including inadequate caloric intake, increased energy expenditure, inappropriate absorption/assimilation, and/or genetic syndromes, all of which have been documented as potential reasons for growth failure in the literature. Energy intake per kilogram of body weight has been correlated significantly to a CHD patient's WFA standard deviation score ($r=0.55$, $p<0.01$ [62]; $r=0.78$, $p<0.001$ [63]) and daily weight gain ($r=0.703$ [64]; $r=0.53$, $p<0.01$ [65]; $r=0.78$, $p=0.003$ [66]). In children with unrepaired defects, energy intake has been reported to be 70%-93% of age appropriate levels (9, 30, 62) at amounts ranging from 46-107 kcal/kg actual weight and 82 kcal/kg ideal weight for height (67, 68). Up to 75% of patients in individual studies may consume less than recommended amounts of energy (27, 30), although some authors report seemingly appropriate voluntary caloric intakes (ie, 115-117 kcal/kg/day of actual weight) (64). However, many experts agree that, while infants and small children with CHD consume calorie levels that might seem reasonable for their actual weight or age, their intake is in fact inadequate when comparing it to their expected (or ideal) weight for current height or when considering energy needs for catch-up growth in those who are already malnourished (34, 35, 67). Furthermore, these infants may have a relative calorie deficit not only because of suboptimal intake, but because of an increase in total energy expenditure per kilogram (32, 34, 69-71), due to increased sympathetic nervous system activity (3, 72-76), increased hematopoietic tissue resulting in polycythemia (attempting to compensate for poor perfusion and cyanosis) (73, 74, 76), increased work of breathing (3, 77, 78), cardiac hypertrophy (3, 73, 74), and/or recurrent infections (61, 73, 74). In one study of 18 infants with ventricular septal defect, energy available for growth was 3.2 kcal/kg in the CHD infants and was 25.2 kcal/kg in the control infants (32); "residual calories" are directly related to weight gain ($r=0.703$, $p<0.001$) (79).

Genetic syndromes, such as trisomy 21, trisomy 13, trisomy 18, Turner syndrome, Williams syndrome, Noonan syndrome, and DiGeorge syndrome (3, 35, 61, 80), are present in up to 5-8% of children with CHD (81), and were actually present in 20% of infants in this particular group of patients. These syndromes are known to produce growth abnormalities irrespective of CHD (82-86); as such, children with CHD as part of a constellation of signs and symptoms characterizing a syndrome may have growth failure that is independent of the heart defect. Infants with genetic syndromes are often excluded from studies of growth and intake in infants with CHD, because the genetic syndrome itself is a confounding variable affecting growth. However, these infants were not excluded in this retrospective study in order to obtain a "real-world" snapshot of all infants with CHD in the postoperative period.

Schwalbe-Terilli (87) and Boctor (45) both conducted retrospective chart reviews examining postoperative nutrition practices and outcomes in heterogeneous groups of CHD infants. First enteral feeding was provided on postoperative days 3 and 5 respectively, compared to day 2 in this study population. The difference in days to first enteral feeding in this study's institution probably represents a continued focus on early enteral feeding to improve clinical outcomes in the ICU population. However, despite enteral feeding three days earlier than infants in Boctor's study, ICU and total hospital LOS in this study were longer (5 days and 13 days vs 4 days and 11 days, respectively). Both other studies reported a worsening in nutrition status during the hospitalization for cardiac surgery, which is in line with data from the current study. Schwalbe-Terilli reported a median weight change of -20 g per day of enteral feeding, and Boctor reported an average change of -11 g per day once transferred from the ICU to regular ward care. Interestingly, patients in this study demonstrated a net gain (median 60 g total increase during the hospitalization). However, 33.8% had a weight loss during the

hospitalization, whereas Boctor reported a 64% incidence of weight loss during the say. Despite a net positive weight change, the current study patients still demonstrated a significant worsening of WFA z-scores during the hospitalization.

Deterioration of nutrition status in the postoperative period after cardiac surgery is due to a mismatch between energy needs and energy intake. Energy needs are higher in these patients after surgery and is even higher in those who have just undergone CBP (88). This is likely because of the significant inflammatory response induced by CPB. CPB is thought to induce systemic inflammation by several mechanisms, including contact of blood with artificial surfaces (the tubing and other equipment used in the CPB circuit) and ischemia-reperfusion injury that occurs when a patient is rewarmed after surgery (89-93). Activation of the acute phase response (marked by an increase in C-reactive protein) and production of pro-inflammatory cytokines, such as interleukin (IL)-6, IL-8 and tumor necrosis factor α, has been documented extensively in children undergoing cardiac surgery with CPB (89, 90, 93-101); activation of these cascades may be an explanation for a transient increase in energy demands. Furthermore, there is a plethora of data showing significant cumulative energy deficits in hospitalized CHD and pediatric ICU patients (88, 102-105); in some studies, CHD patients never reach their goal calorie levels prior to discharge (45, 106). Energy intake is limited because of fluid restriction, multiple periods of *nil per os* (NPO), poor oral intake postoperatively (and at baseline), and gastrointestinal morbidity.

Many infants and children in the immediate postoperative period are fluid restricted (7, 104), which limits the amount of energy and nutrients they can take in. In the ICU, the fluid allowance can be quickly consumed by medications that are crucial to maintain sedation, pain control, paralysis, and cardiac output. Even intravenous nutrition may be withheld if adequate

fluid is not allowed to deliver a reasonable amount of calories. Nutrition intake (particularly enteral feeds) can be limited if procedures such as intubation, extubation, cardiac catheterizations, or operations require NPO status for safety reasons (87, 104). A study of 55 pediatric ICU admissions in which cardiovascular medications were required revealed that feedings were interrupted a mean of 2.8 times per patient, with the most common reason being airway management (107). In our study, enteral feeds were held for a median of eight hours per patient, although 16 patients had enteral feeds held for over 100 hours and one of those patients had feeds held for over 2000 hours. Even though these patients may have been receiving parenteral nutrition during the time in which enteral feeds were held, they were missing out on the immunological and physiological benefits of enteral nutrition.

Almost half of the patients in this study were able to take oral feeds as part of their first enteral nutrition postoperatively. However, feeding morbidity has the potential to significantly impact adequacy of oral feedings. In fact, three infants in this sample were initially allowed to feed orally *ad libitum*, but failed this method of feeding due to inadequate intake, and subsequently required nasogastric feeding. In this study, it was not feasible to thoroughly examine reasons for feeding morbidity, although it is known that reasons for poor oral intake in the postoperative period include poor appetite, oral aversion, suboptimal suck and swallow skills, or laryngeal dysfunction. From a physiologic standpoint, many infants appear to have anorexia or an overall lack of interest in eating (7, 61, 72, 73, 78, 80), which could be a result of diuretic side effects (3, 35, 72) or cardiac decompensation in those with heart failure (35, 60, 73). Decreased gastric volume due to ascites or hepatomegaly (particularly in children in heart failure) can lead to reduced appetite and early satiety (3, 35, 36, 73, 74, 77, 78, 108), as can poor gastric motility resulting from compromised cardiac output (3, 35, 36, 108). Infants with

tachypnea may have difficulty feeding because of an inability to coordinate suck, swallow, and breathe with rapid respiratory rates. After extubation following a significant duration of mechanical ventilation, it is common for infants to exhibit oral aversion. Prolonged intubation has been noted to be an independent predictor of feeding difficulty (109-111) and days to full oral feeds (112) after CHD surgery in neonates. Infants who have had periods of poor central nervous system perfusion during cardiorespiratory arrest events or operative procedures (113) may have neurological impairment that affects suck and swallow coordination and feeding ability (13, 82). In a study of 33 complex CHD patients in the postoperative period, Skinner and colleagues (114) found that 21% had poor or absent suck mechanism and 9% had poor swallowing coordination. Vocal cord impairment can also result in dysphagia, poor oral intake, and tracheal aspiration (115) and has been shown to be an independent risk factor for feeding morbidity in the CHD patient (111). Vocal cord dysfunction occurs in 1.7% to 44% of CHD patients in other studies (115, 116) and was found to be 3.1% in this particular study cohort. Vocal cord impairment can be a result of irritation from an endotracheal tube or can be due to trauma to the recurrent laryngeal nerve during cardiothoracic surgery (3, 111, 115, 117). Indeed, prolonged intubation greater than seven days has been identified as an independent of predictor of dysphagia (OR=74.7, p=0.001) (116), presumably due to effects on laryngeal function. It is important to note, however, that not all infants and children with vocal cord dysfunction have dysphagia or aspiration, although studies are not consistent on this point (114, 115). While many infants improve in oral feeding skills significantly throughout their postoperative course, some require long-term nutrition support. Current studies report that 13%-45% of infants are discharged to home with a nasogastric or gastrostomy tube to supplement oral intake (45, 46,

106, 110); in this study, 15.4% of infants required supplementation with tube feeds, either via nasogastric or gastrostomy tubes, at discharge.

Perceived and actual gastrointestinal intolerance are major barriers to adequate enteral nutrition in the ICU population (87, 107, 118), and signs and symptoms of intolerance, such as abdominal distention, diarrhea, and vomiting, are more common in post-cardiac surgery patients than in other types of patients in the pediatric ICU, partly because of the medications used to maintain cardiac output and sedation; these drugs are known to affect gastric motility (119). NEC has also been documented in CHD cohorts, even though it is historically thought to be a condition primarily confined to the premature infant. Between 1.62% and 13% of term infants with CHD are diagnosed with the condition (120-124), compared to 0.017% in the general term infant population (125). In this study, 1.5% of infants developed NEC during the postoperative period. NEC is the most common intra-abdominal/gastrointestinal emergency of infancy (17), in the most advanced cases, resulting in bowel perforation, peritonitis and death. The pathogenesis of NEC in CHD is largely unknown, although it is hypothesized that poor cardiac output and hypothermia associated with CPB result in poor gut perfusion and mesenteric ischemia (126). Gut flora proliferate with enteral substrate and invade the injured bowel wall. Subsequently, proinflammatory cytokines are released, resulting in systemic inflammation and bowel necrosis (127). NEC affects nutrition status, as treatment generally includes cessation of enteral feeds, in addition to other medical and/or surgical interventions; if significant bowel resection is required, infants can live with the sequelae of NEC for many years after resolution of the initial insult.

Hypoplastic Left Heart Syndrome After Stage I Palliation

Because of the heterogeneity of this sample with regards to type of cardiac lesion, type of surgical intervention, and patient age, it is difficult to intensively examine trends among specific

subgroups. However, given the relatively large number (n=20) of infants with HLHS after stage I palliation, it was possible to look at feeding practices and outcomes in these specific infants and compare those data to what is available in the current literature surrounding postoperative nutrition for infants with HLHS. These infants are considered to be particularly high risk due to their cardiac physiology and requirement for a three-staged surgical approach for palliation, and as such, have been studied by several authors, all trying to identify strategies to improve nutrition-related outcomes in these patients after the first surgical procedure (the Norwood procedure).

Weight at admission (3.1-3.5 kg) in other HLHS studies (42, 51, 53) was similar to the mean weight in this study's patients (3.01 kg). Kelleher and colleagues (53) reported a mean WFA z-score at admission of -0.4, which was in line with the current mean z-score of -0.46. Admission length measurements (49 cm) were similar to those reported by Braudis et al (51) (49.3-50 cm).

In the postoperative period, the HLHS patients required parenteral nutrition for a median duration of 176.5 hours, which is in contrast to 51-116 hours reported by Braudis, and 6 days (144 hours) reported by Kelleher, and Pillo-Blocka (50). Enteral feeds were started on postoperative day 4.2, which is similar to that reported by Braudis and Pillo-Blocka (day 4) and Davis (128) (day 4.8), but less than that reported by Del Castillo (52) (day 5.5-7.5), Jeffries (42) (day 5.9), or Weiss (129) (day 5-8). It is conceivable that, while these infants were started on enteral feeds earlier than those in other studies, the rates were started and/or advanced conservatively, allowing for a significant overlap between parenteral and enteral nutrition, as enteral nutrition advanced and as parenteral nutrition was weaned off and finally discontinued. This could explain a longer parenteral nutrition duration in the current study infants.

Rates of complications in the postoperative period were generally in line with or less than other reports. NEC incidence was 10% in this study, and ranged from 0% in one subgroup in Braudis' study to 27% in one subgroup of Del Castillo's study. Laryngeal dysfunction was reported in only one other study by Davis, and was 18.5% versus 5% in the current study. Death was significantly less, with only 5% mortality, compared to 11%-19%, 13%, and 32% reported by Del Castillo, Weiss, and Pillo-Blocka, respectively. In theory, complication rates could be due to differing acuities in the differing studies, although surrogate measures of critical illness, including lowest pH, highest lactate level, and duration of ventilation, did not appear to be drastically different in any of the studies compared.

ICU LOS in the current study was 12 days, which is similar to what was reported by Braudis (10-12 days) and Kelleher (13 days). Total hospital length of stay, however, was a median of 23.32 days, compared to 16-19 days reported by Braudis. Total LOS was more similar for Del Castillo's infants (21.5-28 days) and Kelleher's subjects (21 days). Time to reach goal enteral (oral or tube) feeds is difficult to compare among studies, since each study defined full feeds differently. For instance, Braudis and colleagues chose 108 kcal/kg, Jeffries et al chose 100 kcal/kg (the value chosen for this study), and Pillo-Blocka used 120 kcal/kg. Jeffries et al observed that their infants required 11.1 days to reach goal enteral feeds of 100 kcal/kg, whereas infants in this study only required 7.5 days. Pillo-Blocka and Braudis both reported net decreases in body weight at discharge compared to admission, whereas in this data set, a net positive increase of 145 grams was noted for the entire hospitalization. It is crucial to point out here, however, that despite the increase in weight, discharge WFA z-score still worsened significantly in these patients from -0.46 to -0.85, owing to an average daily weight gain of 7.2 g/day during

the hospitalization, which is less than one-third of recommended weight gain for infants less than three months of age.

While there are inherent challenges in comparing outcomes from studies of HLHS infants done at different centers, it can be beneficial to review these papers to glean "best practices" that could be used to optimize nutrition management of HLHS and other at-risk CHD infants. One practice emerging from these papers is the use of a standardized approach to enteral feeding in postoperative CHD infants, and more specifically in those status post the Norwood procedure. Many centers across the country have instituted feeding algorithms, protocols, or guidelines to act as decision aids in helping physicians, nurses, and dietitians manage enteral feeding for these nutritionally fragile infants. At Children's Hospital Boston, Braudis et al reported a reduction in number of hours of parenteral nutrition (116 hours versus 51 hours, $p=0.03$) and a trend towards statistical significance in reduction of ICU LOS (19 days versus 16 days, $p=0.07$), total hospital LOS (12 days versus 10 days, $p=0.07$), and incidence of NEC (11% vs 0%, $p=0.07$) after institution of an enteral feeding algorithm in HLHS infants. Additionally, Del Castillo reported a reduction in the incidence of NEC (27% to 6.5%, $p<0.01$) and severity of NEC after implementation of an enteral protocol at Children's Hospital Los Angeles. The clinical utility of enteral feeding algorithms and decision aids has been well-recognized beyond the postoperative CHD infant. Other nutritionally fragile pediatric populations, including premature infants, have benefited from feeding with this approach via reduction in rates of NEC, reduced dependence on parenteral nutrition, earlier achievement of full enteral feeds, and improvements in nutrition status (93, 130, 131). Benefits have also been noted in the pediatric and adult intensive care unit populations, with a reduction in the time to goal enteral feeds (132-134) and an increased

delivery of nutrients from enteral nutrition (135-139), with little to no change in adverse events, like gastrointestinal complaints or tracheal aspiration (132).

Recommendations

The author's institution does not currently have a standardized approach to enteral nutrition management for HLHS and other CHD infants in the postoperative period. Such an algorithm or clinical guideline could outline indications for beginning enteral feeds, how to begin and advance feedings, when and how to transition from tube feeds to oral feeds, and how to manage perceived gastrointestinal tolerance. Furthermore, this sort of algorithm could guide the clinician on timing of consultation of subspecialists, such as Pediatric Speech-Language Pathology, Pediatric Gastroenterology, or Pediatric Surgery for feeding therapy, persistent feeding intolerance, or evaluation for gastrostomy tube placement, respectively. A draft algorithm incorporating these elements is available in Appendix C.

Despite the allure of improved outcomes, there are challenges to algorithm creation and implementation that must be considered. Perhaps one of the biggest downfalls of their use in clinical practice is physician non-adherence to the protocol (137). As such, algorithms must be easy to use and understand and should be evidence-based to improve clinician adherence. Furthermore, algorithm implementation does not end with creation of the decision tool; rather, staff communication, education, and reminders are important for ongoing algorithm utilization. The algorithm must evaluated for efficacy in improving outcomes after it has been implemented, and must be updated regularly with the newest evidence and best practices available in the medical literature (136, 137, 140).

Limitations

As mentioned, the heterogeneity of the study group limits conclusions that can be drawn about particular types of defects or surgical procedures; this approach was chosen, however, in order to improve the total number of available study subjects who could be examined to explore opportunities for improvement in nutrition management. Another major limitation is the retrospective nature of this study. The chart review approach allowed some data points to be very easily collected on a relatively large sample size; however, it was difficult to delve deeper to determine specific etiologies for issues like feeding morbidity or cessation of enteral feeds. This could potentially be addressed by using a prospective study design for future examinations of nutrition outcomes in similar study groups. Similarly, future data analysis could center on nutrition outcomes and obstacles in CHD infants whose nutrition management varied by a predetermined factor, such as initial feeding caloric density, to determine "best practices" that could be incorporated into a feeding algorithm. The retrospective nature of the study also prevents optimal monitoring of accuracy of anthropometric measurements. While nursing staff endeavor to be as accurate as possible in weighing and measuring children, a variety of factors can impede this goal, including patient acuity, fluid overload, parent refusal of measurement, or different measurement tools. Ideally, measurements would be taken by trained, consistent study personnel using the same scales and length boards, using a rigorous predetermine procedure.

Conclusion

In conclusion, this study demonstrated that a heterogeneous group of 100 post-surgical CHD infants exhibited some element of impaired nutrition status (WFA z-score) at birth, which continued to deteriorate in the time period between birth and palliative or corrective surgery and then again in the time period between surgery and discharge from the hospital. Decreasing WFA

z-score is likely due to a multitude of reasons that overall contribute to an imbalance between energy intake and energy expenditure both pre and postoperatively. Outcomes in HLHS infants were, overall, similar in this study as compared to others, although areas for improvement were identified. Strategies such as implementation of an enteral feeding algorithm for these high-risk infants can be beneficial in terms of reducing the risk of NEC and the dependence on parenteral nutrition. A standardized approach to enteral nutrition could be a viable opportunity to improve nutrition-related outcomes in HLHS and other CHD infants at the author's institution, and this paper could serve as control data to which post-algorithm nutrition outcomes data could be compared.

APPENDICES

APPENDIX A

COMMON CONGENITAL HEART DEFECTS

Defect name	**Defining features**
Acyanotic defects	
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
Cyanotic defects	
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

Used with permission from reference 141.

APPENDIX B

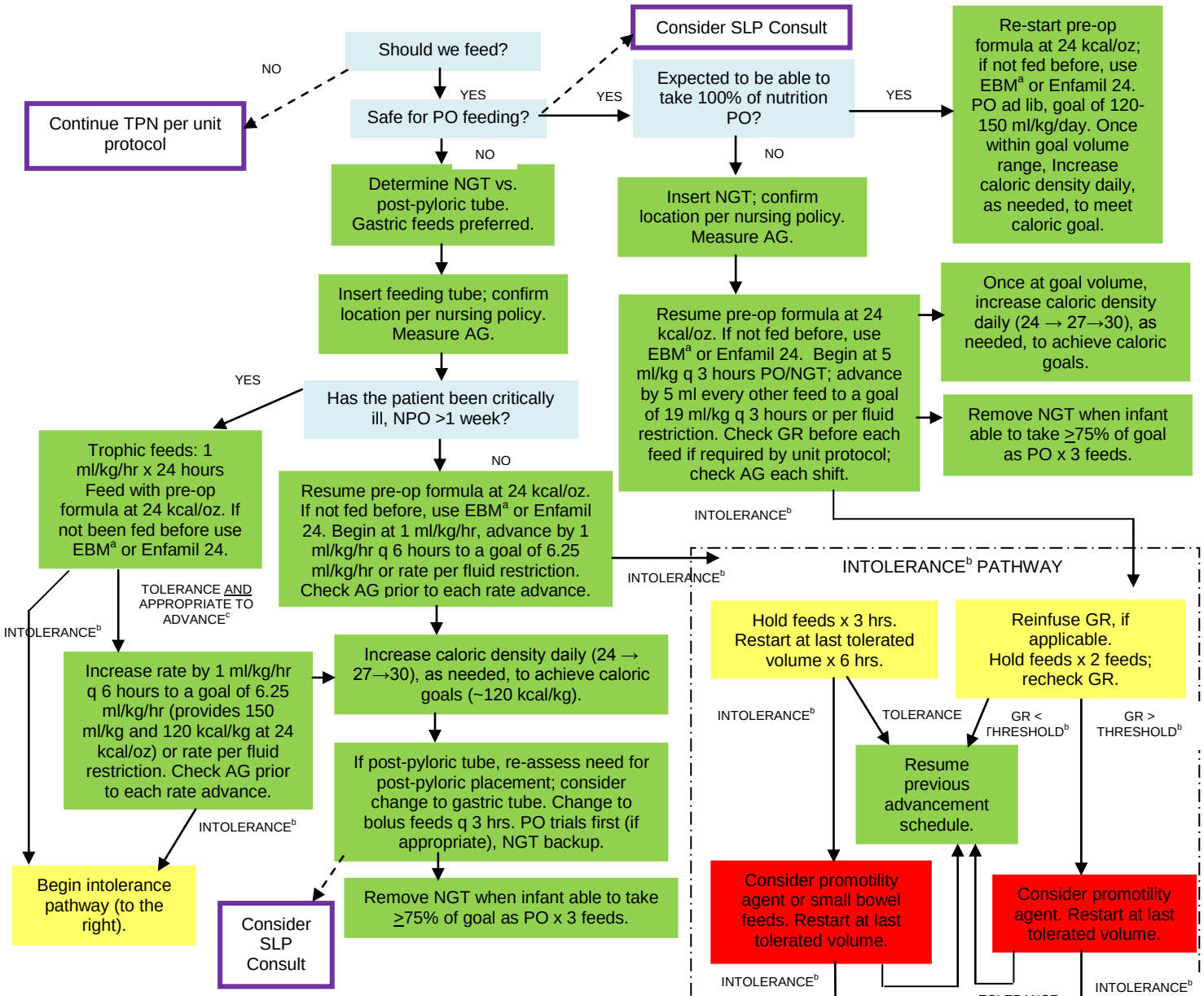
COMMON SURGICAL PROCEDURES TO PALLIATE OR CORRECT CONGENITAL HEART DEFECTS

Procedure	**Description**
Arterial switch	The pulmonary artery and aorta are switched so they arise from the correct ventricle (pulmonary artery from the right ventricle and aorta from the left ventricle).
Atrioventricular canal repair	The ventricular and atrial septal defects are repaired using a patch made from synthetic material. Valve malformations are also repaired.
Bi-directional Glenn procedure	The superior vena cava is anastomosed to the right pulmonary artery to allow passive flow of unoxygenated blood from the top half of the body to the lungs, bypassing the right side of the heart. This is usually the second stage of palliation for infants with a single ventricle.
Blalock-Taussig (BT) shunt	A connection between a branch of the aorta and the pulmonary artery using a patient's own vessel (classic version) or synthetic material (modified version); improves pulmonary blood flow in patients with intracardiac mixing of oxygenated and unoxygenated blood.
Central shunt	Similar to the BT shunt, except that the connection occurs between the aorta and the pulmonary artery (at the area where the pulmonary artery divides to form the left and right pulmonary arteries).
Coarctation of the aorta repair	The narrowed portion of the aorta is excised and the two ends are reconnected.
Damus-Kaye-Stansel procedure	The aorta and pulmonary area are joined together; this procedure is generally done in single ventricle infants where the defect is something other than hypoplastic left heart syndrome.
Hybrid procedure	A stent is placed to keep the ductus arteriosus open, and bands are placed on each branch pulmonary artery to restrict blood flow.
Norwood with modified BT shunt	The pulmonary artery is sacrificed to construct a new aorta for infants with hypoplastic left heart syndrome; pulmonary blood flow is then maintained by a BT shunt.
Norwood with Sano modification	The pulmonary artery is sacrificed to construct a new aorta for infants with a hypoplastic aorta; pulmonary blood flow is then maintained by a right ventricle to pulmonary artery conduit (Sano shunt).
Rastelli procedure	A patch is used to repair a ventricular septal defect; an artificial conduit is used to route blood from the right ventricle to the pulmonary artery.
Septal defect repair (atrial or ventricular)	A patch or stiches are used to close the defect in the wall that separates the atria and/or ventricles.
Tetralogy of Fallot repair	The pulmonary artery stenosis is repaired by removing excess tissue, and the ventricular septal defect is repaired using a patch.
Total anomalous venous return repair	The pulmonary veins are reattached to the left atrium in the correct position.

Information obtained from reference 142.

APPENDIX C

FEEDING ALGORITHM FOR POSTOPERATIVE INFANTS WITH CONGENITAL HEART DEFECTS



[a] Fortify EBM when minimum volumes are conducive to fortification.

[b] Signs/symptoms of feeding intolerance:

- Gastric residual ≥ 1/3 of single feed volume x 2 consecutive feeds.
- Increase in abdominal girth by ≥ 10%
- Vomiting or diarrhea
- Blood in stool (if blood in stool, switch feeds to Pregestimil 20 and resume last tolerated rate).
- Consider NEC workup for persistent/severe GI symptoms.

[c] If clinically appropriate, leave infants who are critically ill (ECMO, very high pressor use, etc.) on trophic feeds.

Abbreviations

AG = abdominal girth	GR = gastric residual
NGT = nasogastric	PO = per os (oral)
q = every	SLP = Speech Language Pathology
TPN = total parenteral nutrition	

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