Variation in growth of infants with a single ventricle

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Abstract

Objective

Growth failure is common among infants with a single ventricle and is associated with adverse surgical outcomes. The National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC) registry captures data about the care and outcome of infants with hypoplastic left heart syndrome (HLHS). Our objective was to evaluate interstage growth variation among sites caring for infants with HLHS and to identify nutritional practices common among sites achieving best growth outcomes.

Study Design

This was a retrospective analysis of infants in the NPC-QIC registry who had presented for their superior cavopulmonary anastomosis (SCPC) and whose surgical site had enrolled $\geq$4 eligible patients in the registry. *Primary outcome variable*: weight-for-age z-score (WAZ) change between Norwood discharge and presentation for SCPC (interstage period). Blinded, structured interviews were performed with each site regarding site-specific nutritional practices. Practices common among sites with positive interstage WAZ changes were identified.

Results

Sixteen centers enrolled 132 infants from 12/2008-12/2010. Median age at SCPC: 5 months (2.6 to 12.6). Median interstage WAZ change: -0.29 (-3.2 to 2.3). Significant variation in WAZ changes among sites was demonstrated ($p<0.001$). Sites that used standard feeding evaluation prior to Norwood discharge and that closely monitored for specific weight gain/loss red flags in the interstage period demonstrated significantly better patient growth than those that did not use these practices ($p=0.002$).

Conclusions

Considerable variation exists in interstage growth among patients receiving care at these 16 surgical sites. Standardization of interstage nutritional management with focus on best nutritional practices may lead to improved growth in this high-risk population of infants.

Introduction

Staged surgical palliation of infants with hypoplastic left heart syndrome (HLHS) has led to dramatic improvements in survival in a medically fragile population. This process begins with the Norwood operation, and the ultimate goal is a Fontan-type circulation.[1-3] Institutional variability and learning curves contributed significantly to early operative and peri-operative mortality, however, post-Norwood survival has improved considerably in the recent era.[4-7] The interstage period, the time between hospital discharge following the Norwood procedure and presentation for the volume-unloading superior cavopulmonary connection (SCPC), typically performed at 4 to 6 months of age, remains a high-risk time for this population, as mortality across many centers approximates 10%.[8] The small number of patients with HLHS cared for at each surgical center has limited the gathering of evidence-based practices in this population.

Adequate growth and nutrition are critically important in infants with HLHS. Poor growth and nutrition are common in infants with congenital heart disease, and malnutrition among patients with complex congenital heart disease, such as HLHS, has been estimated to be as high as 53%.[9] Poor nutrition in children with complex congenital heart disease is associated with infection risk, increased hospital stay and mortality following cardiac surgery.[10-12] Recent studies have suggested that a strict regimen of interstage monitoring, including daily weight checks, results in improved survival among patients with HLHS.[13, 14] Nonetheless, there remains considerable variation in outpatient interstage nutrition management and monitoring.[8] Large multi-center studies evaluating the efficacy of specific practices have not been performed.

The National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC) is the first multicenter quality improvement collaborative within pediatric cardiology.[15] The mission of the NPC-QIC is to improve the outcomes of care for children with congenital heart disease through a national quality improvement collaborative network of multidisciplinary clinical teams and families, working together to collect longitudinal data and conduct quality improvement research intended to accelerate the development and transition of new knowledge into practice. The collaborative is a nationwide network currently comprising 44 pediatric cardiology centers in North America that contribute data to maintain a registry of infants with HLHS. This collaborative provides the infrastructure to better define best practices and outcomes in this relatively rare condition.

The purpose of this study was to describe growth patterns of infants with HLHS enrolled in the NPC-QIC collaborative. We sought to describe variation in growth patterns between surgical centers and to identify specific nutritional practices that were associated with centers with better growth outcomes.

Methods

Study Design

This was a retrospective analysis of patients enrolled in the NPC-QIC registry who had presented for their SCPC. In order to be enrolled in NPC-QIC registry, patients are required to be discharged home following their Norwood operation. Families consented to be enrolled in the registry and enrollment was approved by individual institutional review boards at all participating sites. For this study, patients from the NPC-QIC registry were included from surgical sites who had enrolled ≥4 patients who had undergone SCPC. No patients who met inclusion criteria were excluded from analysis. Study data were collected from two sources: 1) Patient information that had previously been entered into the NPC-QIC registry database, and 2) Structured interviews carried out with members of each surgical site that qualified for inclusion in the study.

Data collection

Demographic Data

Demographic data collected included gender, gestational age, race, and age at presentation for SCPC.

Anthropometric measures

Antrhopometric measures included absolute weight and weight for age z-scores (WAZ) at the following three discrete time points: initial neonatal hospital admission, discharge following Norwood and admission for SCPC. The change in WAZ between Norwood discharge and SCPC admission was calculated for each patient. Zero change in WAZ was considered adequate growth, a WAZ change > 0 was considered positive growth, and a WAZ change < 0 was considered negative growth. WAZ scores were calculated using the Centers for Disease Control Epi-Info™ program (Centers for Disease Control, Atlanta GA).

Medical and Surgical Management

Information regarding medical and surgical management prior to the SCPC was recorded, and included initial cardiac diagnosis, type of Norwood procedure performed and hospital course following the Norwood procedure. Medications prescribed in the interstage period were recorded, as well as the use of home monitoring systems using pulse oximetry and/or home scales.

Nutritional management

Detailed information was recorded regarding each site's nutritional practices before and following the Norwood procedure through hospital discharge, and during the interstage period leading to the SCPC. Specifically, information regarding caloric goals, mode of feeding, use of inpatient and outpatient nutritional specialists, and modalities of monitoring for growth failure were documented. In order to obtain more detailed information on these practices, structured interviews were performed with individuals from each center regarding center-specific nutritional practices. Each site was asked to identify an individual or group that was most knowledgeable about their center's nutritional practices. Each individual or group then participated in a structured phone interview regarding these practices. Two investigators (JA, MH) carried out all of the interviews. The interviews included both multiple choice and open-ended questions regarding the nutritional practices at each site. A third investigator (KV) was present during all of the interviews and recorded the answers to the questions. The interviewers were blinded to the identity of those being interviewed.

Statistical methods

The *primary outcome variable* was WAZ change between neonatal discharge following the Norwood procedure and admission for the SCPC. Variation in WAZ changes within surgical sites and among sites was identified. Sites were then divided into two groups: those with a positive median interstage WAZ change (indicating better growth) among their patients and those with a negative median interstage WAZ change (indicating poorer growth) among their patients. Patient demographics and site practices between these two groups were compared. Nutritional practices were identified that were common to those sites with a positive median interstage WAZ change among their patients.

All statistical analyses were performed using SAS version 9.2 (SAS Institute Inc., Cary, NC). To measure potential differences in patient demographics between the two groups (i.e. sites with positive versus negative median interstage WAZ), mixed linear models were used with each demographic variable as the dependent variable, group as the independent variable, and site as a random effect to account for the clustering of patients within each site. Next, the association of each nutritional intervention and the two groups was measured; Fisher's exact test was used to determine statistical significance.

To determine the effect that each nutritional intervention had on WAZ, a linear regression model was run with the median change in WAZ for each site as the dependent variable and the presence (X=1) or absence (X=0) of the nutritional intervention as the independent variable. The slope from this model was used to estimate change in the median WAZ for a site if the nutritional intervention was practiced. All statistical tests were conducted at the two-sided, 5% level of significance.

Results

Demographic Data

One hundred fifty-three infants in the NPC-QIC registry presented for SCPC between 12/2008 and 12/2010. One hundred thirty-two infants were identified from 16 different sites that had enrolled $\geq$ 4 patients; these patients were included for analysis. Patient characteristics are shown in **Table 1**. Cardiac diagnoses are shown in **Figure 1**.

Medical and Surgical Management

All patients had a stage 1 surgical palliation. A Norwood with Blaylock-Taussig shunt was performed in 30 patients (23%); a Norwood with right ventricle to pulmonary artery conduit was performed in 86 patients (65%); a hybrid stage 1 procedure was performed in 10 patients (8%) and a Damus-Kaye-Stansel procedure with Blalock-Taussig shunt was performed in 6 patients (4%). All but 2 of the stage 1 hybrid procedures were performed at a single surgical site. Following Norwood the median intubation time was 5 days (1-54) and the median hospital length of stay was 31 days (9-126).

Nutritional management

Following Norwood, the median time to full enteral feeds was 13 days (4-77). By the time of discharge, 83% of the infants were being fed orally, 46% were being supported with a nasogastric or nasojejunal tube, and 8% had a gastrostomy tube in place. The majority of patients were fed with human milk formula with 40% being fed breast milk. The median caloric content of the formula or breast milk was 24 kcal per ounce (20-30). At the time of discharge 26% of the patients were prescribed an H2 blocking medication and 23% were prescribed metoclopramide.

Interviews regarding specific nutritional practices revealed that 94% of sites in this study utilize an inpatient dietician in making nutritional decisions during the Norwood hospitalization. During the Norwood post-operative period, 50% of the sites used a standard feeding evaluation to determine whether infants could orally feed adequately. This most commonly was accomplished using speech therapy consultation. When making decisions to advance post-operative feeding, 38% had a protocol in place to help advance enteral feedings and 31% standardly used either an H2 blocker or metoclopramide post-operatively. At the time of discharge following Norwood, 63% of sites sent their patients home with scales to monitor daily weight and weight changes. Similarly, 63% of sites used specific "red flags" to monitor for early signs of growth failure with 9 sites engaging in both of these practices. These specific "red flags" included failure to gain 20 grams per day over a 3 day period or weight loss >30 grams per day on any day in a 3 day period, as used by Ghanayem *et. al.* in their study of home surveillance in infants with a single ventricle.[13] At cardiology outpatient visits during the interstage period 69% of the sites had a dietician available for consultation regarding changes in nutritional management. Dieticians standardly evaluated patients at each interstage cardiology visit at 12% of the sites, and were available to see the patients as needed at 57% of the sites. Between clinic visits 69% of the sites made regular phone contact with families regarding caloric intake and growth. These phone calls varied between once every other week to multiple times per week depending on the clinical situation.

Outcome measures

As can be seen in **Figure 2**, there was significant variation among sites in change in WAZ during the interstage period in patients cared for at each of the surgical sites ($p<0.0001$). Seven sites had a median interstage WAZ change >0 and 9 sites had a median interstage WAZ change < 0. **Table 2** demonstrates patient and management characteristics compared between sites that had a positive median interstage WAZ change and those that had a negative median interstage WAZ change. There were no patient characteristics that differed significantly between sites with positive WAZ changes and sites with negative WAZ changes. The only management difference between groups was the performance of stage 1 hybrid procedures, performed almost exclusively at one of the surgical sites.

Compared to sites with negative WAZ changes, sites with positive WAZ changes more commonly used *standard evaluation of infants' feeding ability* following the Norwood (83% vs. 30%), used *home scales for interstage weight monitoring* (86% vs. 50%), made *regular phone contact with families* during the interstage period regarding nutrition and growth (100% vs. 50%) and used *specific weight gain/loss "red flags"* to identify patients with growth failure in the interstage period (100% vs. 40%). Patients from sites who standardly evaluated feeding abilities prior to Norwood discharge and that used specific weight gain/loss red flags in the interstage period had significantly better interstage weight gain than centers who did not use these practices ($p=0.001$). The benefit on growth of several different nutritional practices is shown in **Figure 3**. Individually, utilization of each of these practices had a varying effect on the change in interstage WAZ. We looked at combinations of these individual practices to find the most significant "bundle" of nutritional interventions in affecting interstage weight gain. The combination of standard post-operative feeding evaluation before Norwood discharge and close weight monitoring in the interstage period with the use of home scales and specific weight gain/loss red flags resulted in the greatest effect, with an increase of change in WAZ of 0.98, compared to sites that did not use these monitoring interventions. Several factors seemed to have little effect on interstage WAZ changes, including feeding modality and the use of standard gastrointestinal medications such as H2 blockers, proton pump inhibitors or promotility agents.

Discussion

We found considerable variation not only in the interstage growth among sites, but also in the nutritional care processes individual sites use. We also identified practices common to sites with better growth outcomes. Addressing variation in practices may be the first step in improving outcomes in this high-risk group of patients. Improvements in surgical technique and medical management over the past decades have led to improvement in clinical outcomes in even the most complex of congenital heart disease.[7] Yet, despite these improvements there continues to be significant variation in management practices among individuals and institutions caring for children with congenital heart disease.[16-18] It has long been demonstrated in other industries that reduction in variation leads to safer practices, cost reduction and improved quality.[19, 20] Interventions made in healthcare that decrease variation in management have shown similar results including improved clinical outcomes, safer practices, and cost reduction.[21-23] Standardization of healthcare practices reduces process variation and provides a foundation on which new approaches can be tested more effectively. Monitoring for post-operative feeding difficulties as well as growth problems in the interstage period are areas where standardization of practices may lead to improvement in outcome.

We found that the most significant factors associated with better growth between centers were related to evaluation of feeding ability during the Norwood post-operative period as well as the nature of monitoring for growth failure in the interstage period. Results of standard evaluation of ability to feed following Norwood surgical palliation in infants with a single ventricle have been reported in previous single center experiences.[24, 25] Following Norwood palliation patients may experience vocal cord paralysis or paresis and may also have oral-motor dysfunction, both of which may delay advancement of caloric intake.[25] It has been shown that use of a post-operative feeding algorithm following Norwood leads to improved post-operative survival, decreased incidence of necrotizing enterocolitis and decreased time to reaching goal caloric intake.[24, 26] We postulate that standard evaluation of oral-motor feeding abilities following Norwood surgery allows for identification of patients with feeding problems early, eliminating any delay in diagnosing these problems.

Similar to infants with other chronic illnesses, there may be an assumption among those that care for patients with HLHS that these children simply are unable to grow normally. This study reveals that assumption is not likely true. Sites that followed growth of these infants more closely, by using specific red flags for growth failure requiring weight checks at minimum every three days, generally had better growth outcomes. This would suggest that attention to early signs of growth failure can facilitate changes in nutritional management to potentially correct this deviation. In order to follow growth every few days, sites need resources in place to allow families to gather weights and also to report them back to their caregivers. In addition to a data gathering and reporting system, there must be someone with nutrition expertise available to monitor growth closely in these infants, and intervene when necessary with nutritional changes. Sites that use a home monitoring system, specific weight gain red flags, and make regular contact with families regarding feeding and growth are able respond to early signs of growth failure.

While this study was able to look for the first time at growth patterns between multiple surgical centers in infants with a single ventricle, it is limited by its retrospective nature. Only patients who were eligible and consented were enrolled in the NPC-QIC registry. There may have been clinical differences between patients enrolled in the registry and those who refused to consent. The NPC-QIC registry includes site and patient specific data deemed important to understanding the clinical course of this high-risk group of infants and to offer a foundation for starting to reduce variation and perform quality improvement work. The registry has limited information on specific nutritional practices, which forced us to perform structured interviews with sites regarding their practices. While the interviews were carried out with those at each site most expert in the site's nutritional practices, individual patient charts were not reviewed. Therefore, it is unknown whether these nutritional practices were applied uniformly to all patients at each site. Future work in this area will include applying these best nutritional practices across centers and monitoring for changes in individual patient growth after establishing new practices.

The findings in this study were made possible because of the cooperation among multiple clinical centers involved in the NPC-QIC. When caring for infants and children with rare conditions it is difficult to determine best practices because of the limited number of patients an individual provider or center cares for. As we seek to find and implement practices that will lead to improved outcomes in our patients it is imperative that we cooperate and share practices and outcomes among caregivers through patient registries and collaborative groups. The importance of these activities has been recognized by the American Board of Pediatrics. In fact, requirements for maintenance of certification now emphasize assessing quality of care and demonstrating systematic improvement of care for children, a requirement that can be met by involvement in these types of groups.[27]

Conclusion

Variation exists in the nutritional practices and growth patterns of infants with a single ventricle, when these factors are compared across surgical centers. Centers with better growth performance tend to use nutritional practices that allow early identification of feeding difficulties and very close monitoring for early signs of growth problems. Understanding this variation and the suggestion of best growth monitoring practices is the first step in standardizing nutritional monitoring and moving toward elimination of growth failure in this high-risk group of infants. The involvement in national collaboratives and registries makes it possible to answer clinical questions about patients with uncommon disease processes.

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Figure 1: Cardiac diagnoses

Key Figure 1: HLHS: hypoplastic left heart syndrome; MA: mitral atresia; AA: aortic atresia; MS: mitral stenosis; AS: aortic stenosis; DILV: double inlet left ventricle; DIRV: double inlet right ventricle; CAVC: complete atrioventricular canal defect; DORV: double outlet right ventricle

Figure 2: Change in WAZ between S1 discharge and S2

Key Figure 2: WAZ: weight-for-age z-score; S1: stage 1 surgical palliation; S2: stage 2 surgical palliation

Figure 3: Effect of specific nutritional interventions on interstage WAZ changes

Key Figure 3: WAZ: weight-for-age z-score; NG: nasogastric; NJ: nasojejunal; GI: gastrointestinal