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ORIGINAL ARTICLE

Home Monitoring Program Reduces Mortality in High-Risk Sociodemographic Single-Ventricle Patients

Daniel Alexander Castellanos[1] · Cynthia Herrington[2,4] · Stacey Adler[2] · Karen Haas[2] · S. Ram Kumar[2,4,5] · Grace C. Kung[3,5]

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Abstract A clinician-driven home monitoring program can improve interstage outcomes in single-ventricle patients. Sociodemographic factors have been independently associated with mortality in interstage patients. We hypothesized that even in a population with high-risk sociodemographic characteristics, a home monitoring program is effective in reducing interstage mortality. We defined interstage period as the time period between discharge following Norwood palliation and second-stage surgery. We reviewed the charts of patients for the three-year period before (group 1) and after (group 2) implementation of the home monitoring program. Clinical variables around Norwood palliation, during the interstage period, and at the time of second-stage surgery were analyzed. There were 74 patients in group 1 and 52 in group 2. 59 % patients were Hispanic, and 84 % lived in neighborhoods where over 5 % families lived below poverty line. There was no significant difference in pre-Norwood variables, Norwood discharge variables, age at second surgery, or outcomes at second surgery. There were more Sano shunts performed at the Norwood procedure as the source of pulmonary blood flow in group 2 (p value <0.05). There were more unplanned hospital admissions and percutaneous re-interventions in group 2. Patients in group 2 whose admission criteria included desaturation had a 45 % likelihood of having an unplanned re-intervention. Group 2 noted an 80 % relative reduction in interstage mortality ($p < 0.01$). In a multiple regression analysis, after accounting for ethnicity, socio-economic status, and source of pulmonary blood flow, enrollment in a home monitoring program independently predicted improved interstage survival ($p < 0.01$). A clinician-driven home monitoring program reduces interstage mortality even when the majority of patients has high-risk sociodemographic characteristics.

Keywords Interstage · Home monitoring program · Single ventricle · Sociodemographic · Social · Demographics

Abbreviations

IS	Interstage
SVR	Single-ventricle reconstruction
CHLA	Children's Hospital Los Angeles
BTS	Blalock-Taussig shunt
NPC-QIC	National pediatric cardiology-quality improvement collaborative

Introduction

In a population-based study, the prevalence of critical congenital heart disease was found to be 15.6 per 10,000 live births and stillbirths [1]. Many of the diagnoses classified under the term 'critical congenital heart disease' include single-ventricle anatomy and physiology. The

✉ Daniel Alexander Castellanos
dcastell@alumni.nd.edu

1 Department of Pediatrics, Children's Hospital Los Angeles, 4650 Sunset Blvd., Mailstop #68, Los Angeles, CA 90027, USA

2 Division of Cardiothoracic Surgery, Children's Hospital Los Angeles, 4650 Sunset Blvd., Mailstop #66, Los Angeles, CA 90027, USA

3 Division of Cardiology, Children's Hospital Los Angeles, 4650 Sunset Blvd. MS #34, Los Angeles, CA 90027, USA

4 Department of Surgery, Keck School of Medicine of University of Southern California, Los Angeles, CA, USA

5 Department of Pediatrics, Keck School of Medicine of University of Southern California, Los Angeles, CA 90027, USA

underlying cardiac defects in single-ventricle physiology can be quite variable, ranging from atrioventricular or semilunar valve atresia with unbalanced ventricles to hypoplastic single-ventricle anatomy. Surgical palliation usually involves three operative procedures leading to Fontan circulation.

In cases where the initial palliation is a Norwood-type procedure, the period between discharge after Norwood and admission for the second stage, Glenn or superior cavopulmonary anastomosis, is commonly referred to as the interstage (IS) period. This period has been observed to be a high-risk time for sudden death [2]. Published data from the Single Ventricle Reconstruction (SVR) Trial estimate IS mortality at 2–20 % [3]. Due to the high risk of mortality in the IS period, many institutions have developed an IS monitoring program. Whereas its exact nature may vary from institution to institution, almost universally, an IS program involves home monitoring by the parents with a pulse oximeter and scale to measure weights. There is often frequent contact between clinical practitioners and the family either with additional outpatient visits or phone calls. Prior studies have demonstrated a significant effect of IS clinics in reducing mortality [2, 4, 5].

The SVR trial identified patient-specific factors that were associated with increased mortality after Norwood procedure. Two crucial sociodemographic variables identified were ethnicity and living in an area with >5 % population below poverty level [3]. The impact of an IS monitoring program in such a high-risk sociodemographic population has not yet been evaluated. The purpose of our current study is to evaluate the role of the IS home monitoring program at Children's Hospital Los Angeles in reducing IS mortality in patients undergoing Norwood palliation at our center, with specific attention to high-risk sociodemographic variables.

Patients and Methods

This retrospective, single institution review was approved by the Institutional Review Board of Children's Hospital Los Angeles (CHLA). All patient information was extracted from the CHLA cardiothoracic surgical database and electronic medical records.

Patient Population and Study Design

All infants undergoing Norwood procedure at CHLA and who were discharged home before their second-stage palliation were eligible for this study. The IS program was formally instituted at CHLA in January 2010. Patients who underwent a Norwood procedure for the first 3 years after institution of the IS program, i.e., between January 2010 and October 2012, (group 2) were compared to patients who underwent Norwood in the immediate preceding three-year period from January 2007 to January 2010 (group 1). Exclusion criteria included infants who transferred care to another facility after Norwood, infants whose IS care was conducted by cardiologists in geographically distant areas, and patients discharged to hospice following Norwood.

Patients in group 1 received the standard of care at that time, which included scheduled follow-up with their primary cardiologists. Patients in group 2 were discharged home with a pulse oximeter and weight scale. Spot pulse oximeter readings were recorded three times a day, and weights recorded daily. This group of patients had weekly telephone calls from IS nurse specialists, apart from scheduled follow-up visits with their primary cardiologists. Patients were triaged for follow-up based on clinical status reported during telephone calls or cardiologist visit. High-risk criteria that resulted in patients being sent to the emergency department included: If the surgical incision area shows signs of infection or separation, child has a fever higher than 101.5 °F, child demonstrates increase in cyanotic appearance, child has had more than one episode of emesis or diarrhea in a day, child has increased respiratory effort, problems taking two feeds in a row, sleepiness, fussiness and inconsolability, or oxygen saturations persistently below 75 %. In the absence of these high-risk criteria, patients had routine biweekly appointments with our IS specialists. Patients were also seen more frequently in IS clinic if there was noted difficulty with weight gain or per their cardiologist's request.

Data Collection and Analysis

Data were collected through retrospective chart review. Patient demographics, IS re-admission and survival data, planned and unplanned re-intervention details and variables around Glenn procedure were collected and tabulated. Data on the percentage of residents living below the poverty level in a census block were obtained by publicly available data published by the US Census Bureau through the Web site Social Explorer. Statistical analysis was performed using SAS 9.4 with univariate and multiple regression analyses. p value <0.05 was considered significant.

Results

There were a total of 74 patients in group 1 and 52 patients in group 2. There was no significant difference in any patient characteristic prior to Norwood operation. Table 1 displays patient characteristics prior to IS period.

Sociodemographic Variables

We specifically focused on sociodemographic factors that are associated with higher IS mortality. To this end, family self-reported data on ethnicity were collected. Also, based on the address provided by the families, the percentage of families living below poverty level in their domiciliary area was identified. In group 1, 62 % of the families identified themselves as Hispanic and 54 % identified themselves as Hispanic in group 2. Group 1 had 87 % of families living in a census block with more than 5 % living below the poverty level, while group 2 had 79 %. There was no significant difference with regards to these variables between groups 1 and 2.

Variables at Norwood Procedure

There was no significant difference in median age at the Norwood procedure. In keeping with the publication of the SVR trial results, there was a significant difference ($p < 0.05$) in the source of pulmonary blood flow between groups 1 and 2. In group 1, 45 % of patients received a Blalock-Taussig shunt (BTS), 54 % of patients received a Sano shunt (right ventricle to pulmonary artery conduit), and 1 % of patients underwent a hybrid procedure. In group 2, 13 % of patients received a BTS, 87 % of patients received a Sano shunt, and no patient underwent a hybrid procedure.

Interstage Characteristics

We observed differences in several characteristics in the IS period following institution of IS monitoring. There was a trend toward increased weight gain in patients in group 2 in the IS period—average weight gain per day increased from 16 gm/day in group 1 to 20 gm/day in group 2 (Table 2). This improvement in weight gain was materialized without a significant difference in the number of patients who had supplemental feeding via enteral tube (20 % in the group 1 and 29 % in group 2). There was also a trend toward increased use of supplemental oxygen in group 2 (45 % patients compared to 28 % in group 1).

There was a 35 % increase in unplanned hospital re-admissions in the IS period in group 2 compared to group 1 (p value <0.05). There was also a 78 % increase in percutaneous intervention in group 2 (48 vs. 27 %, $p < 0.01$). As noted in the Methods section, we have developed intentionally inclusive and broad criteria that triggered emergency department visit and/or admission to the hospital for work-up of IS patients. We investigated which trigger for admission was most likely to result in an unplanned re-intervention. There were 40 re-admissions in group 2 triggered by 54 high-risk criteria (some patients met multiple criteria). Of the 33 re-admissions that included hypoxia as an admitting symptom (cyanosis noted by parents, low saturation on routine monitoring, or respiratory distress), there were 14 (42 %) unplanned interventions and one hospital mortality. The re-interventions included 12 percutaneous interventions and three surgical interventions (one patient had both a surgical and percutaneous intervention). Nine patients were admitted with gastrointestinal issues (failure to feed, vomiting, or diarrhea) of whom one (11 %) had unplanned percutaneous re-intervention. Nine re-admissions had fever as an admitting symptom, of which one (11 %) needed percutaneous re-intervention. Three patients were admitted with dysrhythmias, and one (33 %) needed cardioversion. We then investigated predictors of need for re-intervention. As expected, use of a Sano shunt correlated with an increased need for re-intervention on univariate analysis. On multiple

Table 1 Patient characteristics prior to interstage period

	Group 1	Group 2
Number of patients	74	52
Male	58 %	60 %
Hispanic	62 %	54 %
Greater than 5 % of families living below poverty level	87 %	79 %
Median age at Norwood surgery (days)	4	5
Median weight at Norwood surgery (kg)	3.2	3.2
Median Norwood length of stay (days)	32	36
Median weight at Norwood discharge (kg)	3.3	3.4
Source of pulmonary blood flow after norwood*		
BT shunt	45 %	13 %
Sano	54 %	87 %
Hybrid	1 %	0 %

* Statistically significant, p value <0.05

Table 2 Patient characteristics during the interstage period

	Group 1	Group 2
Median time between norwood discharge and presentation for stage II (days)	158	139
Percentage of patients with at least one interstage readmission*	57 %	77 %
Percentage of patients undergoing at least one percutaneous intervention*	27 %	48 %
Average weight gain/day during the interstage period (gm/d)	16	20
Percentage of patients with supplemental oxygen at presentation to stage II	28 %	45 %
Percentage of patients undergoing g-tube placement prior to presentation for stage II	20 %	29 %
Median age at stage 2 (days)	205	190
Median Glenn length of stay (days)	8	13

* Statistically significant, p value <0.05

regression analysis, after accounting for source of pulmonary blood flow and sociodemographic factors, enrollment in IS program was independently associated with need for re-intervention in the IS period.

The most significant benefit we observed after institution of an IS monitoring program was an 80 % relative risk in IS mortality (Fig. 1, 18.9 % IS mortality in group 1 vs. 3.8 % in group 2, $p < 0.001$). On univariate analysis, presence of genetic syndrome, birth weight, ethnicity, and enrollment in IS clinic were associated with mortality. We constructed multiple regression models to identify predictors of IS mortality. Genetic syndrome (OR 3.8, 1.6–6.9) and enrollment in IS program (OR 0.6, 0.4–0.8) were independently associated with mortality. After accounting for use of a Sano shunt, socioeconomic status and ethnicity, enrollment in IS program persisted as an independent negative predictor of IS mortality.

Glenn Characteristics

There was no statistically significant difference in median age at Glenn procedure (Table 2). There was also no difference in need for concomitant procedures during Glenn, or hospital length of stay following Glenn.

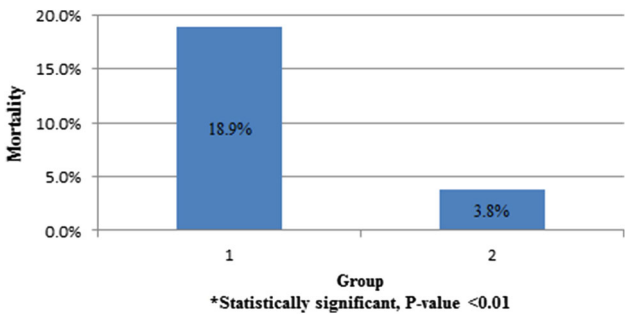

Fig. 1 Interstage mortality in group 1 and 2. There was an 80 % relative reduction in interstage mortality in group 2 (p value <0.01)

Discussion

The interstage period between discharge after Norwood and admission for Glenn has classically been a time of high mortality in patients with single-ventricle physiology. Several prior studies have investigated variables associated with IS mortality [3, 6, 7]. In the multivariable modeling of the SVR trial data, gestational age <37 weeks, Hispanic ethnicity, living in a census block with 5.4–13 % of residents below the poverty level, presence of aortic and mitral atresia, modified BTS in subjects with postoperative no/mild Aortic valve regurgitation, and greater number of post-Norwood complications were all predictors of IS mortality [3]. Given the magnitude of IS mortality, several centers began implementing a home monitoring program [2, 4, 5, 8, 9] with the goal of improving IS outcomes. Prior reports have demonstrated salutary reduction in IS mortality following institution of such IS programs [2, 4, 5]. This has resulted in a favorable shift in practice pattern such that the proportion of patients enrolled in IS surveillance increased from 33 % in a 2012 study [10] to 83 % in a recent National pediatric cardiology-quality improvement collaborative (NPC-QIC) report [11]. Overall IS mortality rate reported from national aggregate data (regardless of implementation of a high-risk clinic) has significantly decreased to 9 % according to the 2014 NPC-QIC study. Some studies, on the other hand, have not been able to demonstrate an improvement in IS mortality with IS monitoring [8]. In particular, patients with high-risk anatomic substrates, such as heterotaxy patients, have no improvement in mortality despite intense IS follow-up [7]. No study to date has evaluated the effect of IS clinic on other high-risk populations identified by the SVR trial.

Due in part to our geographic location, our center caters to a disproportionate number of patients of Hispanic race and from low socioeconomic status—both of which are independently associated with increased IS mortality [3].

Given this unique patient population, our study focused on the effects of home monitoring program on IS mortality in this vulnerable socioeconomic cohort. Enrollment in an IS program was associated with a 15 % absolute reduction in mortality after accounting for differences in clinical variables and sociodemographic factors. Although the exact cause of increased mortality in specific socioeconomic groups cannot be discerned, it has been suggested that limited access to medical care and limited family awareness of clinical deterioration may contribute to adverse outcomes in these patients. Home monitoring programs such as ours implement specific protocols for communication between families and providers and also increase access to specialty care avenues. As such, these programs have the potential to significantly reduce mortality particularly in these high-risk sociodemographic groups.

Our IS program includes routine monitoring of systemic saturations and weight gain. In addition, by design our program has a more liberal policy of admitting 'borderline' patients—a policy designed to better fit the patient population seen at our center. Along these lines, we observed a 35 % increase in unplanned re-admission in group 2 patients. A prior report by Petit et al. [8] did not show any difference in rates of unplanned readmission between groups before and after implementation of a home monitoring program. Interestingly, an analysis by Cross et al. [12] of NPC-QIC data identified unplanned readmissions as an IS mortality risk factor. In the absence of a focused IS surveillance program, unplanned re-admissions are likely to include patients with delayed presentations, and missed complications, which would be associated with increased mortality. In our study, increased re-admission was specifically related to an over-cautious policy of liberal admissions, and in fact resulted in reduced mortality. Along with increased re-admission, there was also a significant increase in percutaneous interventions after the implementation of an IS monitoring program. A study by Siehr et al. [5] also showed that following implementation of an IS program, 17 % of all IS readmissions required major interventions. Of the various categories of high-risk criteria used by our IS program for re-admission, hypoxia and related symptoms were most likely to result in an unplanned re-intervention. This would imply that, in our cohort of patients, the highest yield from the IS clinic was in identifying inadequate pulmonary blood flow that manifests as signs and symptoms of low systemic oxygenation. It is further conceivable that although not statistically significant, the 96 % increase in use of supplemental oxygen was also likely in response to the identification of subtle signs of hypoxemia. Collectively, we believe that the IS program is capable of earlier identification of subtle clinical deteriorations that allows more timely re-interventions and avoidance of 'near-miss' IS deaths. The 80 % relative reduction in IS mortality supports this hypothesis.

A major confounding factor in our study was the significant increase in use of Sano shunts for pulmonary blood flow in group 2 patients. The SVR trial has shown an increase in unintended cardiovascular interventions from the time of initial procedure to 12 months of age in patients who receive a Sano shunt [13]. Similarly, our study also showed a correlation between use of Sano and re-intervention on univariate analysis. However, on multiple regression analysis, enrollment in IS program remained an independent predictor or re-intervention, after accounting for type of pulmonary blood flow. Given that patients with a Sano shunt have an increased re-intervention rate, it is tempting to speculate that an IS program may be more beneficial in this subset of patients by identifying the need for re-intervention early. The sample size of our study and the large proportion of Sano shunts preclude such an analysis within this dataset.

On the second parameter followed by IS monitoring, we did not notice a statistically significant difference in weight gain in patients enrolled in IS program compared to those who were not. It has been established that the IS period is a time of poor growth [4]. Lower body weight at discharge after Norwood is associated with increased IS mortality [7], and improved weight gain in IS period resulting in increased weight-for-age *z*-score at stage II is associated with fewer complications during Glenn [11]. There is wide variability in feeding patterns among the various centers to ensure adequate weight gain in the IS period. It has been suggested that feeding route may be an independent predictor of IS mortality [12]. In our center, we have a bias toward percutaneous enteral feeding tubes to ensure adequate nutritional support. There was no significant difference in the number of patients with *G*-tubes in our two groups studied, implying that the difference in weight gain, albeit not statistically significant, may reflect better overall physiologic state of the children undergoing routine monitoring.

Limitations

Limitations in our study included the nonrandomized nature of our retrospective review. This is a single institution study with a limited subset of patients. Given the nature of our study, an era effect (better outcomes with accumulated experience) cannot be excluded, though unlikely. Confounding factors that impacted quality of care (such as the focus on discharge planning) which may have occurred simultaneously with the IS program could have led to the improvement in mortality statistics.

Conclusion

In sum, our study clearly establishes that enrollment in a home monitoring program leads to earlier identification of clinical deterioration, more re-intervention, and a significant improvement in IS survival ($p < 0.01$). Such a reduction in mortality can be achieved even when the majority of enrolled patients have high-risk sociodemographic characteristics. This may serve as a model for outpatient monitoring of other high-risk groups in the pediatric population.

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Compliance with Ethical Standards

Conflict of interest The authors have no conflicts of interest relevant to this article to disclose.

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