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ORIGINAL PAPER

Associations of Home Monitoring Data to Interventional Catheterization for Infants with Recurrent Coarctation of the Aorta and Hypoplastic Left Heart Syndrome

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Abstract

The post-Norwood interstage period for infants with hypoplastic left heart syndrome is a high-risk time with 10–20% of infants having a complication of recurrent coarctation of the aorta (RCoA). Many interstage programs utilize mobile applications allowing caregivers to submit home physiologic data and videos to the clinical team. This study aimed to investigate if caregiver-entered data resulted in earlier identification of patients requiring interventional catheterization for RCoA. Retrospective home monitoring data were extracted from five high-volume Children's High Acuity Monitoring Program®-affiliated centers (defined as contributing > 20 patients to the registry) between 2014 and 2021 after IRB approval. Demographics and caregiver-recorded data evaluated include weight, heart rate (HR), oxygen saturation (SpO_2), video recordings, and 'red flag' concerns prior to interstage readmissions. 27% (44/161) of infants required interventional catheterization for RCoA. In the 7 days prior to readmission, associations with higher odds of RCoA included (mean bootstrap coefficient, [90% CI]) increased number of total recorded videos (1.65, [1.07–2.62]) and days of recorded video (1.62, [1.03–2.59]); increased number of total recorded weights (1.66, [1.09–2.70]) and days of weights (1.56, [1.02–2.44]); increasing mean SpO_2 (1.55, [1.02–2.44]); and increased variation and range of HR (1.59, [1.04–2.51]) and (1.71, [1.10–2.80]), respectively. Interstage patients with RCoA had increased caregiver-entered home monitoring data including weight and video recordings, as well as changes in HR and SpO_2trends. Identifying these items by home monitoring teams may be beneficial in clinical decision-making for evaluation of RCoA in this high-risk population.

Keywords Hypoplastic left heart syndrome · Interstage monitoring · Coarctation of the aorta · Cardiac catheterization

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Introduction

Pediatric cardiac interstage home monitoring programs [1, 2] along with the National Pediatric Cardiology Quality Improvement Collaborative [3] have helped to decrease interstage mortality in infants with hypoplastic left heart syndrome (HLHS). Yet, increased survival rates have shed light on an array of comorbid and significant complications during the interstage period and beyond. One of the most prevalent is hemodynamically significant recurrent coarctation of the aorta (RCoA), occurring with an estimated incidence of 18% for neonates and infants who undergo a Norwood procedure [4]. The clinical presentation of RCoA is highly variable, ranging from asymptomatic to severe, with symptoms, such as feeding intolerance and increased work of breathing. Left untreated, RCoA can lead to declining ventricular function and worsened atrioventricular valve regurgitation (AVVR). Asymptomatic infants are typically diagnosed with RCoA via routine echocardiography. Ideally, this would be discovered before changes in ventricular function or AVVR occur. However, traditional models of monitoring physiologic data during the interstage period are reactive to arising problems and may miss opportunities for early identification of changes associated with RCoA.

Most centers that perform single-ventricle palliation have an interstage home monitoring system connecting parents to care providers. Traditional home monitoring requires caregivers to document weight, oxygen saturation (SpO_2), heart rate (HR), intake and output, and parental concerns in a notebook or binder. These data are sent to care teams at least weekly and when issues arise [5]. As the burden of relaying information is largely placed on the caregivers, communication failures carry the potential for missing important and urgent clinical signs. As a result, many teams are leveraging mobile health technologies to reduce barriers, allowing caregivers to electronically record patient data on devices (e.g., tablets, smartphones) that are nearly ubiquitous and provide that data directly back to the clinical teams. These teams are often led by nurses and nurse practitioners who review data trends and proactively communicate with families on a weekly basis, monitoring for emerging clinical problems [6].

To date, remote home monitoring data have been shown to provide signals indicative of risk for catastrophic events (e.g., cyanosis). However, given the slow rate of change and varied presentation, it remains unknown if these same vital signs and “Red Flag” data can provide similar insight into long-term morbidities, such as the need for intervention during interstage catheterization in patients with RCoA. Clinical red flags and changes indicating RCoA may include upper and lower blood pressure difference, tachypnea, tachycardia, higher oxygen saturations, fluid retention, and an inability to gain weight by the infant. The ability to identify those who will require intervention would allow for improved family counseling prior to appointments and facilitate proactive communication of potential red flags for both frontline staff and families. This is especially important at centers with large catchment areas, as many families travel a significant distance to be seen in clinic.

In order to study the hypothesis that remote home monitoring data acquired during the interstage period will differ significantly in patients with RCoA that require intervention compared to those that do not require intervention, this study utilizes a set longitudinal data from a large, multi-site, interstage remote home monitoring application: Children’s High Acuity Monitoring Program (CHAMP®)—Cardiac. CHAMP has been shown to be associated with significantly fewer unplanned hospitalizations requiring intensive care days, shorter delays in care, lower resource utilization, and lower incidence of interstage growth failure [7]. The app also has a high degree of adherence for data entry—approximately 75% vital sign input by parents—providing an excellent opportunity to understand patient characteristics outside of the hospital setting.

Material and Methods

The study population comprised infants with single-ventricle congenital heart disease who underwent Norwood palliation and were included in the CHAMP® multi-site research registry at participating centers between 2014 and 2021. All infants underwent a cardiac catheterization (including planned pre-Glenn catheterization) which included evaluation of RCoA between their initial Norwood discharge and scheduled bidirectional Glenn procedure. In order to mitigate bias that may have arisen from differences in monitoring or interventional practices at centers with few patients, the study cohort was limited to infants treated at high-volume centers. This was defined as centers contributing at least 20 patients to the CHAMP registry during the study period, in which at least ten patients underwent a Norwood procedure. Infants were excluded if they were never discharged home in the interstage period or had less than one month of data available at the time of data extraction. For infants with multiple cardiac catheterizations, only the first procedure date was used in order to reduce bias in future data collection practices that may arise due to observations or results of earlier interventions. This study was approved by the Children’s Mercy institutional review board with Request-to-Rely approval at the University of Missouri-Kansas City School of Medicine.

Data Source

Data for each infant included clinical factors collected during their neonatal/Norwood admission and interstage home monitoring information, routinely collected through the CHAMP mobile application. CHAMP is a software platform with mobile application that enables nearly instantaneous home monitoring and improves communication during the vulnerable interstage period. Caregiver-acquired physiologic home monitoring data are transferred to a secure cloud-based server which houses a multi-site database registry. The cloud-based server has multiple functions, including sending data to local electronic medical records, automated analytics, and real-time data and alerts to the clinical team for out-of-range data. Since 2014, the CHAMP multi-site registry has grown with use at a multi-site level, covering over 1000 patients living in 25 states with care provided by 12 pediatric hospitals in the USA. Currently, there are nine hospitals using CHAMP for the care of children with heart disease for home monitoring with a 95% consent rate by parents for participation in the cardiac registry.

Clinical Data

In addition to standard demographics, including the distance between home and the tertiary cardiac surgical center, data related to outcomes during interstage monitoring were collected. These included age at Norwood procedure, age at discharge home, type of pulmonary blood flow source [right ventricle (RV) to pulmonary artery (PA) shunt or modified Blalock–Thomas–Taussig (mBTT) shunt], cardiac catheterization prior to discharge, and echocardiogram information at neonatal discharge (aortic arch gradient, ventricular function, and systemic AVVR). The number of scheduled clinic visits was also recorded, as were details around each cardiac catheterization, including the age of the child, nature of the catheterization (scheduled versus unscheduled), and any interventions performed. For the analysis presented in this manuscript, interventions were defined as either balloon or stent angioplasty of the aorta during the catheterization.

Home Monitoring Data

Parental-reported data entered into the CHAMP mobile application were captured. These included weights, oxygen saturations, and heart rates, as well as the submission of video monitoring data (content of videos was not analyzed as part of this work). Additionally, 'Red flags' were captured that indicate concern to the care team. These flags can be automatically generated due to recorded values outside of acceptable thresholds, healthcare team initiated on data review, as well as caregiver submissions that were of concern. 'Red flag' symptoms include significant vomiting or diarrhea and concerns of dehydration, change in feeding tolerance or feeding difficulty, low oxygen saturations (less than 70%), irritability or change in responsiveness, weight loss over 3 days, bloody stools, sweating with feeds, or any change that is new and worrisome to the care providers.

Data cleaning involved removing SpO_2 less than 60% to remove possible entry error. For cases in which multiple single-day measurements were entered for weights, oxygen saturations, and heart rates, the values were averaged prior to use in any of the feature calculations.

These data were used as part of a featured engineering effort to derive the primary study variables, characterizing information home monitoring data collected in the days leading up to an infant's readmission. These included averages, ranges, measures of variance, and derived time series measures of recoded vital signs, as well as measures of parent data entry patterns. A complete listing of derived study features and their definitions can be found in Table 1.

To address the underlying research question of how patterns of home monitoring data may be associated with the need for intervention for RCoA, data for each infant were collected and derived over a 7-day window prior to the catheterization. These were then associated with baseline data collected at discharge and an outcome defined as use of balloon/stent angioplasty during the encounter catheterization.

Given a high degree of collinearity between the study variables, each metric was studied independently using a logistic mixed effects model. Adjusting for sex, Norwood Surgery type (mBTT shunt, RV-PA shunt), ICU discharge day-of-life, patient race, and age in days at readmission with a random intercept added to account for site-level clustering. Both the age at the time of Norwood and age at the time of initial discharge were log transformed to account for heavy right tails. To aid in interpretation and model convergence, values for each metric were standardized (mean: 0, SD: 1).

Analysis was performed across two distinct cohorts. The first cohort represented all infants and focused on modeling the association between data entry patterns and the outcome (e.g., total number of distinct submissions and number of unique days across the 7-day window). However, for metrics that rely on the raw data collected as part of the monitoring process (e.g., the recorded heart rates or SpO_2), a second cohort of individuals was utilized who each recorded data on at least three of the seven days in the window. In contrast to the data entry patterns, in which no data recorded is itself an important feature, this was done in order to reduce bias that may arise due to parents reporting only a few worrisome values or that may arise due to small samples sizes of data across the prior week. In this way, we can more accurately represent the association between the quantitative values of the home monitoring data and outcome rather than latent associations to entry patterns.

In line with the current literature, this work moves away from the use of arbitrary p-value thresholds for each metric. Rather, for each metric a 10,000-iteration bootstrap was performed, in which the model is refit on resampled cohorts to better estimate the directionality and magnitude of association effect size for the respective metric with incidence of interventional catheterization for RCoA. From this distribution the mean and associated confidence intervals were reported. All feature engineering was completed using Python v3.8.8, numpy v1.20.1, tsfresh v0.18, with mixed model analysis completed using R v4.1.0, with the lme4 v1.1-29 (glmer/bootMer) packages [8–11].

Results

In total, 167 infants met inclusion criteria across the five high-volume CHAMP-affiliated hospitals: Children's Mercy Kansas City (n =45), Seattle Children's Hospital (n=31),

Table 1 Derived study features and definitions

Category	Study feature (Applicable data types)	Definition
Data entry	Total days: Vitals Weights Videos	Total number of days over the observation window for which at least 1 data element was charted in the CHAMP mobile app
	Total entries: Vitals Weights Videos	Total number of unique data entries charted in the CHAMP mobile app over the observation window
Clinical events	Clinic visits count	Number of clinic visits in the 30 days prior to readmission
	Red flag count: Any Feeding Breathing	Number of red flags generated by the CHAMP app during observation window. Any represents any type of flag generated by parent, clinical team, or app. Feeding/Breathing represents counts of those flags specifically related to concerns in those domains
Home monitoring data	Mean, SD, Min/Max, Range SpO_2 HR	A set of common descriptive statistics including Arithmetic mean, standard deviation, minimum, maximum (as discrete values), and range (max–min) across the observation window
	Mean Change SpO_2 HR Weight	The average change between subsequent data points across the observation window
	Slope SpO_2 HR	Beta coefficient from linear model fit on the time series of the respective data type (y-value vital sign measurement value, x-value representing the day the value was recorded in the observation window)

Primary Children's Hospital ($n=30$), Cincinnati Children's Hospital Medical Center ($n = 36$), and Cook Children's Medical Center ($n= 25$). Of these, 6 patients were removed as they either underwent surgical intervention for RCoA during their bidirectional Glenn surgery ($n=5$) or were missing data ($n=1$). These subjects were removed to prevent the introduction of bias that would result from marking these individuals as not RCoA when factors such as timing of bidirectional Glenn or institutional preferences may have impacted intervention decisions. Of the remaining 161 infants, 19 patients were found to have undergone multiple catheterizations, for which only data prior to the first catheterization was included for analysis.

The cohorts' demographics and clinical data are shown in Table 2, while the average values for each study variable by cohort can be found in Table 3. Of the 161 total infants included in the analysis, 44 infants (27.3%) received transcatheter RCoA interventions. Of these, 24 of the 44 (54.5%) infants had a planned angioplasty. Those who underwent treatment for RCoA were more likely to have neo-aortic arch obstruction at the time of their discharge echocardiogram. After the cardiac catheterization, 96 infants discharged home, 18 remained inpatient until the Glenn procedure, and four died. A third of the infants who remained inpatient ultimately needed RCoA intervention, while less than a quarter of those discharged home returned for immediate RCoA surgical intervention.

Figure 1 results from the primary analysis of this work, highlighting the mean effect size and associated confidence interval for the association of each study variable with intervention. Given the exploratory nature of the work, all relationships were assessed at 90% confidence to provide a wider set of potentially informative monitoring data for use in follow-up studies aimed at estimating thresholds of concern across associated study variables. As study variables have been standardized, the magnitude of effect sizes is comparable across features. For convenience, coefficients and confidence intervals were exponentiated to represent odds ratios.

In the 7 days before the catheterization, there was increased adherence to data entry, number of videos, weight, average SpO_2, HR range, and HR variability. Specifically, the data adherence metrics regarding video and weight input showed increased odds of requiring RCoA intervention (mean bootstrap coefficient, [90% CI]): increased number of total recorded videos (1.65, [1.07–2.62]), increased number of days of video recordings (1.62, [1.03–2.59]), increased number of total recorded weight measurements

Table 2 Cohort demographics

		No RCoA intervention ($n=117$)	RCoA intervention ($n=44$)	Total ($n=161$)	Missing data
Patient Sex	Female	45 (38.5)	16 (36.4)	61 (37.9)	0
	Male	72 (61.5)	28 (63.6)	100 (62.1)	
Race	Asian	2 (1.7)	0 (0)	2 (1.2)	0
	Black/African American	14 (12.0)	4 (9.1)	18 (11.2)	
	Other	4 (3.4)	4 (9.1)	8 (5.0)	
	White	97 (82.9)	36 (81.8)	133 (82.6)	
Average age of primary caregiver in years		27.5 (5.5)	27.4 (6.3)	27.5 (5.7)	13
Primary cardiac diagnosis	HLHS	82 (70.1)	30 (68.2)	112 (69.6)	0
	Other	35 (29.9)	14 (31.8)	49 (30.4)	
Type of surgery	Norwood with mBTT shunt	42 (35.9)	12 (27.3)	54 (33.5)	0
	Norwood with RV-PA conduit	75 (64.1)	32 (72.7)	107 (66.5)	
Age at Norwood, days		5.9 (3.8)	8.5 (7.5)	6.7 (5.2)	0
Age at Discharge, days		44.3 (28.5)	51.2 (31.9)	46.2 (29.5)	0
Discharge echocardiogram					
Ventricular Function	Mild/moderate	7 (6.0)	4 (9.1)	11 (6.8)	0
	Normal	110 (94.0)	40 (90.9)	150 (93.2)	
Systemic AV Valve Regurgitation	Moderate/severe	14 (12.0)	7 (15.9)	21 (13.0)	0
	None/mild	103 (88.0)	37 (84.1)	140 (87.0)	
Neo-Aortic Arch Obstruction (> 10 mmHg)	No	103 (88.8)	25 (58.1)	128 (80.5)	2
	Yes	13 (11.2)	18 (41.9)	31 (19.5)	
Clinical Evaluation of Distal Aortic Arch Obstruction (> 10 mmHg) via a blood pressure gradient	No	81 (69.2)	38 (88.4)	119 (74.4)	1
	No information available	36 (30.8)	1 (2.3)	37 (23.1)	
	Yes	0 (0)	4 (9.3)	4 (2.5)	
Catheterization Intervention and Type	None	117 (100.0)	0 (0)	117 (72.7)	0
	Planned angioplasty	0 (0)	24 (54.5)	24 (14.9)	
	Unplanned angioplasty	0 (0)	19 (43.2)	19 (11.8)	
	Planned Stent Placement	0 (0)	1 (2.3)	1 (0.6)	
Age at Catheterization, days		107.0 (40.7)	107.1 (38.3)	107.6 (40.0)	0
Surgery Undergone During Readmission After Cath	No	66 (80.5)	30 (93.80)	96 (84.2)	47
	Yes	16 (19.5)	2 (6.3)	18 (15.8)	
Disposition after Catheterization	Death	4 (4.8)	0 (0)	4 (3.4)	43
	Discharged home	67 (80.7)	29 (82.9)	96 (81.4)	
	Hospitalized until Glenn	12 (14.5)	6 (17.1)	18 (15.2)	

AV atrioventricular, *HLHS* hypoplastic left heart syndrome, *mBTT* modified Blalock–Taussig–Thomas, *PA* pulmonary artery, *RcoA* recurrent coarctation of the aorta, *RV* right ventricle.

*Data are presented as N (%) for categorical variables and Mean (Standard deviation) for continuous variables

(1.66, [1.09–2.70]), and increased number of days of weight recordings (1.56, [1.02–2.44]). Additionally, changes in the physiological factors of SpO_2 and HR also demonstrated increased odds of requiring RCoA intervention: increasing mean SpO_2 (1.55, [1.02–2.44]), increased minimum and maximum values of SpO_2 (1.64, [1.06–2.70]) and (1.60, [1.04–2.53]), respectively, increased variation in HR (1.59, [1.04–2.51]), and increased range of HR (1.71, [1.10–2.80]). The range and slope of SpO_2 were not correlated with the odds of intervention, nor were the minimum or maximum values of HR. Furthermore, weight loss, the total number of red flags, or those concerning feeding/breathing concerns were not correlated with RCoA intervention.

Discussion

Infants with HLHS are prone to complications after undergoing the Norwood surgical palliation, with RcoA (Fig. 2) being a high-risk complication [1]. With such variable symptomology, it can be difficult to discern this complication during the interstage period through home monitoring

Table 3 Description of cohort monitoring data: SD—Standard Deviation, Min—Minimum, Max—Maximum, O_2—Oxygen Saturation, HR—Heart Rate, RCoA—Recurrent Coarctation of the Aorta

	Total	No RCoA Intervention	RCoA Intervention
Adherence Metrics Cohort			
n	161	117	44
Clinic Visit Count	1.6 (1.1)	1.6 (1.2)	1.7 (0.9)
Red Flag Count	0.3 (0.7)	0.3 (0.6)	0.6 (1.0)
Red Flag Count	0.2 (0.5)	0.2 (0.4)	0.2 (0.5)
Red Flag Count	0.4 (0.6)	0.4 (0.6)	0.4 (0.6)
Vitals Total	6.2 (9.8)	6.1 (11.3)	6.4 (4.0)
Vitals Days	3.9 (2.3)	3.7 (2.3)	4.5 (2.0)
Videos Total	3.2 (2.8)	2.7 (2.6)	4.6 (2.8)
Videos Days	2.9 (2.4)	2.5 (2.4)	4.0 (2.3)
Weight Total	4.0 (2.3)	3.7 (2.3)	4.6 (2.1)
Weight Days	3.9 (2.2)	3.7 (2.3)	4.5 (2.0)
CHAMP Monitoring Cohort			
n	127	88	39
Mean O_2	80.6 (2.7)	80.4 (2.8)	81.2 (2.4)
SD O_2	1.8 (0.8)	1.7 (0.8)	1.8 (0.6)
Min Val O_2	78.2 (3.1)	77.9 (3.1)	78.8 (2.9)
Max Val O_2	83.2 (3.0)	82.8 (3.2)	83.9 (2.5)
Range O_2	5.0 (2.4)	4.9 (2.4)	5.1 (2.4)
Slope O_2	− 0.01 (0.05)	− 0.01 (0.05)	− 0.01 (0.04)
Mean HR	136.6 (11.7)	137.2 (11.9)	135.2 (11.4)
SD HR	6.5 (3.7)	6.1 (3.5)	7.3 (4.0)
Min Val HR	127.6 (13.7)	128.7 (14.1)	125.1 (12.4)
Max Val HR	145.8 (12.4)	145.9 (12.1)	145.6 (13.2)
Range HR	18.2 (10.1)	17.2 (9.7)	20.5 (10.6)
Slope HR	− 0.01 (0.16)	0.01 (0.11)	− 0.04 (0.23)
Weight Loss	0.8 (0.7)	0.8 (0.7)	0.7 (0.6)

*Data are presented as *N* (%) for categorical variables and Mean (Standard deviation) for continuous variables

data. Caregivers are routinely asked to enter daily vital sign measurements of infants in CHAMP, similar to recommendations in other home monitoring programs. Our team practice is to evaluate metric changes over seven days. Since the early 2000s, the standard data collected by parents included oxygen saturation, feeding concerns, and red flags symptoms [2, 4, 12].

This study is the first to explore the relationship of home monitoring data associated with RCoA in infants with single-ventricle heart disease who have undergone Norwood palliation. Our findings have highlighted several significant associations: interstage RCoA patients were found to have increased numbers of caregiver-entered home monitoring data and notable physiologic changes in HR and SpO_2. With home monitoring programs, data entry is expected to vary from caregiver to caregiver. Results indicate that caregivers' increased adherence to data entry for video and weight recordings was associated with increased odds of requiring catheterization for RCoA. Although challenging to elucidate the exact underlying reason, we postulate that this notable increase in data entry may result from the caregiver noting a deterioration in the patient's clinical condition, thereby increasing adherence to the recommended CHAMP data entry. However, this hypothesis will require further study as it is possible that the increased data entry followed an echocardiogram that showed evolving arch obstruction or due to the increased contact with the clinical care team outside of the app, not captured in this study.

On neonatal hospital discharge, there was a higher frequency of infants with RCoA with reported neo-aortic arch gradients above 10 mmhg by discharge echocardiogram compared with those that did not have an intervention (41.1% vs. 11.9%). While this incidence of peak aortic arch gradient is similar to the previously reported independent risk factor ($p < 0.0001$) associated with RcoA [13], this does not have obvious clinical implications unless this measure has the potential to be a very early indicator of developing RcoA. However, this would need to be further explored as our study was not designed to assess this. An additional future measure that could give

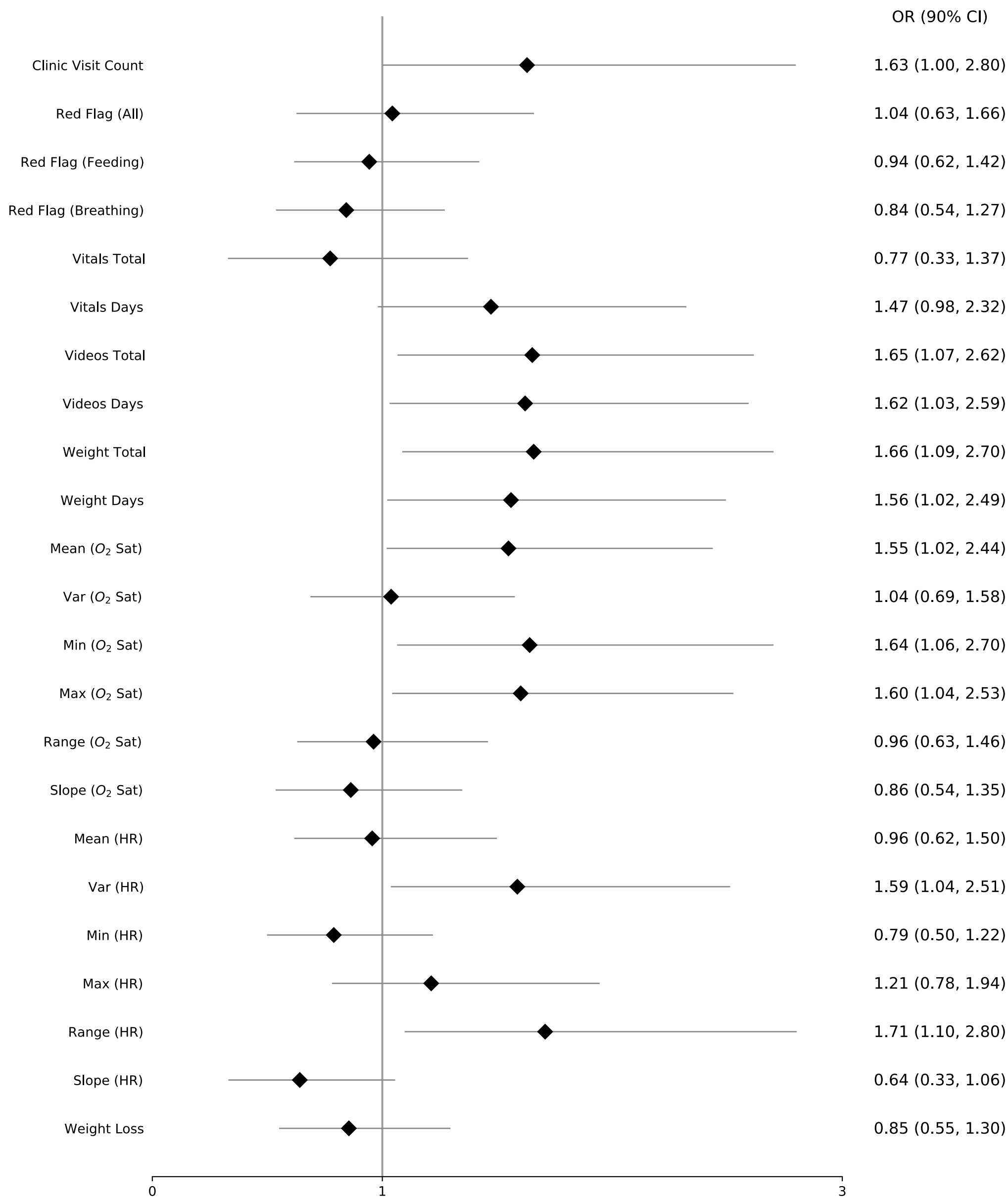

Fig. 1 Association of Study Variables to Intervention During Interstage Catheterization for Patients with RCoA. OR—odds ratio, CI—confidence interval. Point estimates represent the bootstrapped odds ratio and bars represent 90% confidence intervals. Values greater than one can be interpreted as a relative increase in the odds of intervention as the value of the study variable increases, with the magnitude representing the increase in odds relative to a one standard deviation increase in feature magnitude. Conversely for coefficients less than one

additional insight into RCoA is the clinical assessment of blood pressure, especially upper and lower extremity values. While parents do not commonly measure this in the home setting after Norwood palliation, on neonatal discharge there was only a 2.5% incidence of blood pressure gradient of more than 10 mmHg in the cohort. Intermittent interstage clinic assessment of serial blood pressure gradients may provide trends to understanding development along with echocardiographic findings. If noted to have a gradient via echocardiogram or by blood pressure gradient, the healthcare team would likely recommend closer follow-up and the benefit of home monitoring technology; families could supply this information through vital signs, videos, and weight. Mobile health videos can evaluate tachypnea, color, and the overall status of infants with HLHS [14]. Weight data can be assessed for fluid overload or lack of weight gain, which can provide additional information for proactive review by the healthcare team.

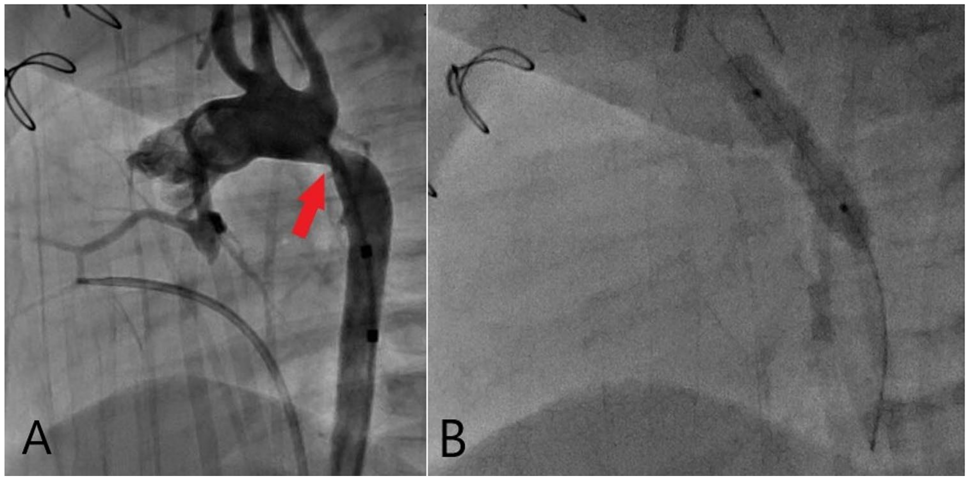

Fig. 2 Recurrent Coarctation of the Aorta and Balloon Angioplasty. **A** Red arrow pointing to significant recurrent coarctation of the aorta. **B** Balloon angioplasty for treatment of recurrent coarctation of the aorta

Aligned with current monitoring guidelines surrounding threshold-based alerts, our findings identified increased mean, minimum, and maximum changes in SpO_2 as associated with increased occurrences of RCoA that required intervention. As part of the Norwood operation, infants undergo placement of an RV to PA shunt or mBTT shunt from the innominate/subclavian artery to the pulmonary arteries. In patients who develop RCoA, there is an increase in resistance to systemic blood flow due to the narrowing of the aortic diameter. As a result, more blood flow preferentially goes to the lungs, which may be evident by increasing oxygen saturation. In some cases, if overall systemic cardiac output decreases as a result of RCoA, worsening ventricular function, or more significant AV valve regurgitation from increased afterload, then a lower systemic venous saturation may offset the increased pulmonary blood flow and correspondingly not result in higher saturations in all patients. Notably, we also found increased variation and range of HR to be associated with increased odds of RCoA intervention. Variability in heart rate in the post-Norwood population has been studied as a part of inpatient ICU care as a possible predictor of upcoming events while on continuous monitoring [15]. However, quantitative thresholds and critical values for changes in vital sign variability have yet to be evaluated in the outpatient remote home monitoring care setting. Although this retrospective observational study was not adequately powered or designed to establish these thresholds, future prospective work could be designed to recommend a level of variability that would indicate need for imaging of the aortic arch. Additionally, the continuous measure of variability itself may be useful, as emergent machine learning risk models may leverage this measure in a way that is not available currently.

Interestingly, red flag concerns by the caregivers were not correlated with increased odds of needing a RCoA intervention. While further study is required, this may result from variation among sites, caregivers, and subjective bias of those individuals in what constitutes a 'red flag.' It should be noted that initial 'red flag' warnings were based on clinical practice without firm data.

Limitations

Typical of observational, retrospective cohorts, this study had several limitations. First, due to the rare nature of HLHS, the sample size for this study was limited. However, as patients were drawn from across five clinical centers and analytical techniques were used to help improve the generalization of estimates, this work provides an important set of potential risk factors for future prospective studies. Second, due to the retrospective nature of the data, we cannot know if adherence increased after a routine echocardiogram showed RCoA and if parents were more compliant while awaiting a scheduled cardiac catheterization to treat the RCoA. Attempts were made to alleviate this issue by evaluating scheduled and unscheduled admissions; however, there were nearly equal numbers of interventions for RCoA in both groups.

Conclusion

The interstage period after the Norwood palliation is a high-risk time for infants with HLHS. Those reviewing home monitoring data should pay close attention to HR range and variability, higher SpO2, sudden increase in weight gain, and higher caregiver adherence to input of home monitoring data. By identifying these trends, knowledge of a high potential for recurrent coarctation may allow for earlier identification of hemodynamic compromise, thereby improving counseling ability for parents prior to appointments.

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Declarations

Competing interests All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

References

1. Ghanayem NS, Hoffman GM, Mussatto KA et al (2003) Home surveillance program prevents interstage mortality after the Norwood procedure. J Thorac Cardiovasc Surg 126(5):1367–1375
2. Hehir DA, Dominguez TE, Ballweg JA et al (2008) Risk factors for interstage death after stage 1 reconstruction of hypoplastic left heart syndrome and variants. J Thorac Cardiovasc Surg 136(1):94-99.e1-e3
3. Schidlow D, Anderson JB, Klitzner TS, JCCHD National Pediatric Cardiology Quality Improvement Collaborative et al (2011) Variation in interstage outpatient care after the Norwood procedure: a report from the Joint Council on Congenital Heart Disease National Quality Improvement Collaborative. Congenit Heart Dis 6(2):98–107
4. Hill KD, Rhodes JF, Aiyagari R et al (2013) Intervention for recoarctation in the single ventricle reconstruction trial: incidence, risk, and outcomes. Circulation 128(9):954–961. https://doi.org/10.1161/CIRCULATIONAHA.112.000488
5. Rudd NA, Frommelt MA, Tweddell JS et al (2014) Improving interstage survival after Norwood operation: outcomes from 10 years of home monitoring. J Thorac Cardiovasc Surg 148(4):1540–1547
6. Shirali G, Erickson L, Apperson J et al (2016) Harnessing teams and technology to improve outcomes in infants with single ventricle. Circ Cardiovasc Qual Outcomes 9(3):303–311. https://doi.org/10.1161/CIRCOUTCOMES.115.002452
7. Bingler M, Erickson LA, Reid KJ et al (2018) Interstage outcomes in infants with single ventricle heart disease comparing home monitoring technology to three-ring binder documentation: a randomized crossover study. World J Pediatr Congenit Heart Surg 9(3):305–314. https://doi.org/10.1177/2150135118762401
8. Harris CR, Millman KJ, van der Walt SJ et al (2020) Array programming with NumPy. Nature 585:357–362. https://doi.org/10.1038/s41586-020-2649-2
9. Christ M, Braun N, Neuffer J, Kempa-Liehr AW (2018) Time series feature extraction on the basis of scalable hypothesis tests (tsfresh–a python package). Neurocomputing 307:72–77
10. Bates D, Mächler M, Bolker B, Walker S (2015) Fitting linear mixed-effects models using lme4. J Stat Softw 67(1):1–48. https://doi.org/10.18637/jss.v067.i01
11. Rudd NA, Ghanayem NS, Hill GD, Lambert LM, Mussatto KA, Nieves JA, Robinson S, Shirali G, Steltzer MM, Uzark K, Pike NA, the American Heart Association Council on Cardiovascular and Stroke Nursing, Council on Lifelong Congenital Heart Disease and Heart Health in the Young, Council on Arteriosclerosis, Thrombosis and Vascular Biology; Council on Clinical Cardiology, Council on Lifestyle and Cardiometabolic Health (2020) Interstage home monitoring for infants with single ventricle heart disease: education and management: a scientific statement from the American heart association. J Am Heart Assoc 9(16):e014548. https://doi.org/10.1161/JAHA.119.014548
12. Whiteside W, Hancock HS, Pasquali SK, Yu S, Armstrong AK, Menchaca A, Hadley A, Hirsch-Romano J (2016) Recurrent coarctation after neonatal univentricular and biventricular norwood-type arch reconstruction. Ann Thorac Surg 102(6):2087–2094. https://doi.org/10.1016/j.athoracsur.2016.04.099
13. Rusin CG, Acosta SI, Vu EL, Ahmed M, Brady KM, Penny DJ (2021) Automated prediction of cardiorespiratory deterioration in patients with single ventricle. J Am Coll Cardiol 77(25):3184–3192. https://doi.org/10.1016/j.jacc.2021.04.072
14. Aly DM, Erickson LA, Hancock H, Apperson JW, Gaddis M, Shirali G, Goudar S (2021) Ability of video telemetry to predict unplanned hospital admissions for single ventricle infants. J Am Heart Assoc 10(16):e020851. https://doi.org/10.1161/JAHA.121.020851
15. Erickson, L.A. (2020). A descriptive correlational study of rate and determinants of parental mHealth adherence to symptom home monitoring for infants with congenital heart disease during the single ventricle interstage period: The DOMAIN study. Published Dissertation in Nursing from the University of Missouri Kansas City via Proquest MoSpace. https://hdl.handle.net/10355/73993

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