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INTRODUCTION

Neonates with single ventricle congenital heart disease (SVCHD) are a complex patient population with a high risk for mortality during the interstage period between stage 1 palliation (S1P) and stage 2 palliation (S2P). During the interstage, patients are either discharged into a home monitoring program, or they remain hospitalized until S2P if they have significant risk factors such as tricuspid valve regurgitation, a persistent oxygen requirement, or a feeding regimen that preclude discharge. Our objective was to retrospectively apply NEONATE scores categorizing patients as high risk (a score>17) or low risk (a score \leq 17) for interstage mortality or transplant established by Dr. Ahmed and colleagues to our home monitoring (safe at home: SAH) cohort, and our patients who remain in the hospital (safe in house: SIH) cohort at different timepoints throughout their clinical course following S1P. The NEONATE score is comprised of the following components: Norwood type (hybrid: 6 points, BTTS: 3 points, Sano: 0 points), ECMO requirement post Norwood (6 points), opiates at discharge (6 points), no digoxin at discharge (9 points), aortic arch obstruction (6 points), tricuspid valve regurgitation ≥ moderate (12 points), and tricuspid regurgitation \geq moderate and an oxygen requirement (28 points).

METHODS

A retrospective analysis of patients with SVCHD with ductal dependent lesions at Children's Medical Center in Dallas, TX was performed. Patients were included if they had undergone S1P with a Norwood with BTTS or Sano followed by S2P or cardiac transplant. They were also included if they underwent a hybrid procedure as their first palliation until either completion of the Norwood BTTS or Sano, completion of S2P, or cardiac transplantation. The patients were divided into two cohorts: patients discharged home into the Safe at Home program, and patients who remained in house following S1P. In order to be included, the patients had to be born no earlier than January of 2017 and they had to complete their S2P or cardiac transplantation by May of 2021. We selected January of 2017 as our starting point in order to account for changes in surgical era at our institution that occurred in 2016. Patients were excluded if they passed away in the immediate postoperative period following S1P, they were born before January of 2017, and/or if they were determined to not be candidates for surgical palliation. Chi-square and Fisher's exact tests were performed to analyze categorical variables, and the Mann Whitney Wilcoxon test was utilized for continuous variables. A p value of <0.05 was considered to be statistically significant

RESULTS

Table 1: The majority of patients in both cohorts had a primary diagnosis of
 hypoplastic left heart syndrome (70% SAH and 76% SIH), were male (77% SAH versus 67% SIH), Caucasian (50% SAH and 48% SIH), were prenatally diagnosed with SVCHD (87% SAH and 81% SIH), did not have genetic syndromes (80% SAH and 86% SIH), and did not have comorbidities (77% SAH and 81% SIH). The mean gestational age and birthweight were 38 weeks and 3.2 kg for the SAH cohort, and 37 weeks and 3.2 kg for the SIH cohort. There was no statistically significant difference in diagnosis, gestational age, birthweight, gender, race, prenatal diagnosis, comorbidities, or presence of genetic syndromes between the two cohorts. SIH patients had a statistically significantly longer duration of intubation post S1P compared with SAH patients. **Figure 1:** The median NEONATE score for the patients discharged into the SAH program was 6 (low risk category).

Figure 2: Seven days post extubation, the SIH cohort had statistically significantly higher NEONATE scores than the SAH cohort (median: 23 versus 9, p value < 0.0001). Median NEONATE scores calculated prior to CVICU transfer to the floor were the following: (SAH: 9 versus SIH: 17, p value: 0.05). Pre-S2P or pre-transplant scores were statistically significantly higher in the SIH cohort (median 15 versus 6, p value: 0.0008). The SAH and SIH cohorts each had one interstage death. There was no statistically significant difference in interstage mortality between the two cohorts (p value: >0.9999).

		Safe at hom (n=30)
Diagnosis		
HLHS		21 (70
Other		9 (30)
DILV		2
Unbalanced AVCD		
defect		1
hypoplasia		4
Critical aortic stenosis		1
Aortic valve atresia,		
transverse aortic arch		1
Gestational age (weeks)		38 (1.9)
Birthweight (kg)		3.2 (0.55)
Comorbidities		
Yes		7 (23%)
No		23 (77%)
Gender		
Male	23 (77%)	
Female	7 (23%)	
Prenatal DX		
Yes	26 (87%)	
No	4 (13%)	
Genetic syndrome/genetic mutation		
Yes	6 (20%)	
No	24 (80%)	
Race		
Caucasian	15 (50%)	
Hispanic	9 (30%)	
Other	6 (20%)	
Duration of initial intubation (days)		
		8 (6)

Figure 3: In the SAH cohort, 93% of patients had NEONATE scores in the low risk category at the time of discharge. Figure 4: 96% of SAH patients with scores <12 remained alive post S2P and transplant free (1 patient died post Glenn). 75% of patients with scores between 12-17 remained alive post S2P and transplant free (1 patient died post Glenn followed by transplant). Both patients who were discharged with scores in the high risk category ultimately died (one died during the interstage, and one died after Glenn followed by ventricular assist device implantation).



Figure 3: NEONATE scores prior to discharge for the SAH cohort

Evaluation of NEONATE Scores and Outcomes Among Patients with Single Ventricle Congenital Heart Disease who either enter the Safe at Home Program or Remain in the Hospital following Stage 1 Palliation prior to Stage 2 Palliation or Cardiac Transplantation.

> Gianna M. Romano^{\perp}, MD, MPH, Thomas M. Zellers, MD^{\perp} Division of Pediatric Cardiology, ²UT Southwestern Medical Center, ¹Children's Health, Dallas, Texas, USA





Figure 1: Box and whisker plot displaying NEONATE scores at discharge for the SAH cohort.



Table 1: Baseline characteristics of the SAH and SIH patients. Categorical variables presented as frequencies (%), and continuous variables presented as a mean (standard deviation).

Figure 2: Box and whisker plots comparing NEONATE scores for the SAH and SIH cohorts at 3 timepoints during their clinical course.



Figure 4: NEONATE scores prior to discharge for the SAH cohort and outcomes.

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Figure 5: Pre-S2P or pre-transplant NEONATE scores for the SAH and SIH cohorts. Figure 6: Pre-S2P and pre-transplant NEONATE scores and the percentage of SAH and SIH patients who survived s/p S2P and remained transplant free

> **Figure 6**: 96%(23/24) of SAH patients with low risk scores and 58% (7/12) SIH patients with low risk scores survived to S2P and remained transplant free compared to 60% of SAH patients and 25% of SIH patients with high risk scores.



Figure 5: Pre-S2P or pre-transplant NEONATE

scores: 76% (SAH) versus 25% (SIH) had scores

<12, 7% SAH versus 35% SIH had scores betwee

12-17, and 17% SAH versus 40% SIH had scores

≥18.

Figure 7: Pre-S2P or pre-transplant NEONATE scores and the percentage of SAH and SIH patients who experienced transplant or mortality.

Figure 7: 4% (1/24) SAH patients versus 42% (5/12) SIH patients with low risk scores experienced transplant or mortality compared with 40% of SAH and 75% of SIH patients with high risk scores. 66% of SAH patients (2/3) who died post S2P had high risk scores pre-S2P, and the SAH patient who died during the interstage had consistent high-risk scores during their clinical course. In the SIH cohort, 1 patient died during the interstage (this patient had a NEONATE score in the high risk category 7 days post extubation and never transferred out of the CVICU). Eight additional patients died: 6 post Glenn, 3/6 (50%) had high risk scores pre-S2P, and 2 post transplant (both patients had high risk scores pre-transplant).



Figure 8: CVICU length of stay post S2P or cardiac transplant for SAH and SIH patients.

Figures 8 and 9: The SIH cohort had statistically significantly longer median CVICU LOS post S2P or cardiac transplant: 31 days (25th%: 14, 75th%: 67, compared to the SAH cohort: 5 days (25th%: 2, 75th%: 12), p value < 0.0001 and statistically significantly longer median total LOS post S2P or cardiac transplant: 54 days (25th%: 26, 75th%: 150, compared to the SAH cohort: 15 days (25th%: 9, 75th%: 21), p value < 0.0002.



Figure 9: Total length of stay post S2P or cardiac transplant for SAH and SIH patients.

CONCLUSION

The NEONATE score when applied to SAH and SIH SVCHD patients could be a helpful tool to prospectively guide clinicians with predicting interstage and potentially post Glenn risk of mortality or transplant, with vast potential to improve patient management and outcomes.

REFERENCES

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