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## Predictors and Outcomes of Heart Block During Surgical Stage I Palliation of Single Ventricle Patients: A Report From NPC-QIC

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## Abstract

**Background:** Mortality in single ventricle cohorts remains high with multiple associated factors. The effect of heart block during stage I palliation remains unclear.

**Objectives:** To study patient and surgical risks for heart block and its effect on transplant-free 12-month survival in single ventricle patients.

**Methods:** Patient, surgical, outcome data and heart block status (transient and permanent) were obtained from the NPC-QIC single-ventricle database. Bivariate analysis was performed comparing patients with and without heart block and multivariable modelling used to identify variables associated with block. One year outcomes were analyzed to identify variables associated with lower 12 month transplant-free survival.

**Results:** In total, 1,423 patients were identified, of which 28 (2%) developed heart block (second degree or complete) during their surgical admission. Associated risk factors for block included heterotaxy syndrome (OR 6.4) and atrial flutter/fibrillation (OR 3.8). Patients with heart block had lower 12 month survival though only in patients with complete heart block as opposed to second degree block. At 12 months of age, 39% of heart block patients died and were more likely to experience mortality at 12 months compared to patients without block (OR 4.9, 95% CI 1.4,17.5, p=0.01).

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**Conclusions:** Though rare, complete heart block following stage I palliation represents an additional risk for poor outcomes among this high risk patient population. Heterotaxy syndrome was the most significant risk factor for development of heart block following stage 1 palliation. The role of transient block on outcomes and potential rescue with long term pacing remains unknown and requires additional study.

#### Keywords

Heart block; Hypoplastic left heart syndrome; Pediatric cardiology; Stage I Palliation; Survival

## Introduction

Staged surgical palliation of patients with single ventricle has undergone significant advancement over the past decades, though morbidity and mortality remain significant.(1-3) Although rare in single ventricle patients, concomitant heart block adds complexity and potential risks for pediatric patients with congenital heart disease. The incidence of heart block in large single ventricle populations is poorly defined, but in the literature has been reported in ~ 4-11%.(4,5) Surgical and patient characteristics such as heterotaxy syndrome and the need for additional atrioventricular valve surgery have been postulated mechanistic drivers associated with heart block in single ventricle populations but data remains sparse in large cohorts, particularly surrounding Stage I palliation (S1P).(6-8) Patients with permanent heart block typically require long term ventricular pacing and pacemaker implantation which has been associated with progressive ventricular dysfunction in older age Fontan cohorts.(7,9) In younger single ventricle cohorts, the associated consequences of heart block and the deleterious effects of chronic ventricular pacing is unknown. In recent data from the Pediatric Heart Network and additional small retrospective studies, patients with perioperative heart block have had decreased transplant free survival to Stage II palliation compared to those without heart block, though the mechanisms are unclear and survival past the second stage palliation is unknown.(4,5,10,11) This study seeks to address the current gaps in the literature surrounding the risk and long term outcomes for patients with single ventricle heart disease with post/perioperative heart block following S1P to inform care for this high risk population.

## Methods

The National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC) is a learning health network composed of over 60 pediatric care centers developed to improve outcomes for patients with hypoplastic left heart syndrome (HLHS) and variants. The NPC-QIC includes a voluntary registry with a standard dataset with data definitions, online web-based data entry, and data quality checks. Institutional review board approval and parental consent or waiver of consent is maintained by individual centers participating in the NPC-QIC database. Additional approval was obtained at Cincinnati Children's Hospital Medical Center for this study #2020–0001 (4/22/2020). The research reported in this paper adhered to Helsinki Declaration guidelines. Phase I NPC-QIC data captures patient, surgical and outcome data from birth through the interstage period and admission for Stage II palliation for patients discharged after S1P. Phase II data includes all variables from phase I

and expands upon this data with additional patient data through 12 months of age including all patients deemed to need a S1P, regardless of discharge status.

#### **Study Population**

All patients included in the NPC-QIC with phase II data were included in the initial analysis. To be included in the data set patients must: 1) have HLHS or variant single ventricle disease and 2) need either Norwood procedure (BT shunt, central shunt or RV-PA conduit), Damus-Kaye-Stansel (DKS) or hybrid procedure.

#### Perioperative Stage I Palliation and Interstage Variables

Data were collected for all patients from birth through admission for second stage palliation including variables in the interstage time period. Patient characteristics including sex, race, primary cardiac diagnosis, additional major congenital anomalies, and heterotaxy status were collected. Surgical and perioperative variables were collected including type of surgical palliation, requirement for additional AV valve repair. Post-surgical and interstage variables were collected including patient ventricular function on the echocardiogram prior to discharge, hospital length of stay (LOS) and whether a patient required readmission following discharge from their stage I surgical palliation and prior to their second stage palliation. Patients with heart block were noted during the initial hospital admission for stage I surgical palliation. The database included variables for heart block, including a separate variable for second and third degree heart block, and patients could have both occur during their hospitalization. Data on first degree heart block was not collected. The registry does not denote whether heart block was permanent or transient only that it was present at some point following stage I surgical palliation. Patients with perioperative heart block were defined as having either second or third degree heart block, with additional sub-analysis performed based on heart block type. Separate discrete variables for pacemaker implant during or after stage I surgical admission were collected.

#### 12 Month Patient Outcome Data

Patients equal to or greater than one year from the date of birth were identified and discrete outcome variables are denoted in the database including: survival, death prior to discharge from stage I palliation, interstage death, death following stage II palliation, heart transplantation and "other death". Only patients with this outcome variable at 1 year of age were included in this 12 month analysis and data is censored at 12 months. Peri-operative data including patient age, weight and length of stay were collected in the cohort of patients surviving to 12 months of age.

#### Statistics

Data were examined for feasibility and correctness, and any data point which was not possible, or feasible was set to missing. Bivariate analysis was initially performed to compare patients with and without heart block to identify patient, surgical and outcome differences between the patient cohorts and variables associated with heart block. Additional bivariate analyses were performed to evaluate survival to stage II palliation as well 12 month survival in the sub-cohort of patients 1 year of age or greater. X<sup>2</sup> test or Fisher's exact test

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were used for categorical variables and Student's t test or Wilcoxon rank sums test was used for continuous variables as appropriate. Based on bivariate analyses, separate models for risk of heart block during stage I admission and risk of death prior to 12 months were created. Survival analysis was performed, comparing survival in patients with/without HB, and also comparing pacemaker placement/not in patients having HB. Logrank tests for equality over strata were performed, and Kaplan-Meier curves were plotted for visual display of this analysis. All analyses were performed using SAS Version 9.4 (SAS<sup>®</sup>Institute Inc., Cary, NC, USA).

## Results

#### Patient and Surgical Factors Associated with Heart Block

In total, 1,423 single ventricle patients were identified having undergone stage I palliation within the phase II cohort contained in the NPC-QIC database (July 2008 - August 2020). Heart block was seen in 2% (28 patients) of this cohort. (Table 1) Of the patients with heart block, 14 had second degree block, 12 had complete heart block and 2 patients had both second and complete heart block seen during their admission. In general, patients with heart block were less frequently black/African American, were less likely on digoxin at time of discharge and were older at the time of Stage I discharge (p 0.05). Heart block patients were more likely to have heterotaxy syndrome and atrial flutter and had an increased mortality compared to patients without heart block (p=0.001). Of the 28 patients with heart block, 8 patients (29%) underwent pacemaker implant prior to stage I discharge. Of the 8 patients with heart block who underwent pacemaker implantation, 4 patients had second degree heart block and 4 had complete heart block. Twenty patients with heart block did not have pacemaker implantation including 10 patients with complete heart block. Three patients underwent pacemaker implantation without heart block. One patient had atrial tachycardia and the remaining 2 patients had no additional arrhythmia coding. Pacemaker implantation was more frequent in patients with heart block (8 patients (29%)) vs. those without heart block (3 patients, (0.2%, p=<0.0001)). One additional patient was readmitted in the interstage period for pacemaker implant in each cohort.

Based on bivariate modelling and plausible relationship to heart block, race (black/African American), presence of heterotaxy syndrome and history of atrial flutter/fibrillation were included in the initial multivarible logistic regression modelling for heart block. Heterotaxy syndrome (OR 6.4(1.8–22.7), p=0.005) and history of atrial flutter/fibrillation (OR 3.8(1.1–13.0), p=0.04) were independently associated with heart block during stage I admission while race was not. (Table 2)

#### Associated Outcomes of Patients with Heart Block

In the cohort of patients with heart block, death or transplant prior to Stage II palliation was 39%, with the majority of those deaths occurring prior to stage I discharge (21%). (Table 3) Comparing patients with heart block to those without, patients with heart block were older at the time of stage I discharge (62.4 vs. 38 days, p=0.05) but had similar Stage 1 LOS (p=0.1). Patients with heart block were more likely to be admitted through the interstage period (p=0.003) and have an unanticipated readmission in the interstage period for pacemaker

implantation (4% vs. 0%, p=<0.0001). Patients with heart block were more likely to have an outcome of death (39% vs. 16%, p=0.001) but not transplantation (4% vs. 4%, p=0.9) compared to those patients without heart block. In addition to a lower associated survival, patients with heart block were also more likely to have death prior to discharge from Stage I palliation (21% vs. 9%, p=0.02) than patients without heart block. (Figures 1 and 2) Despite the presence of heart block, no patient in the heart block group was felt to have an ultimate cause of death directly attributable to arrhythmia.

#### Variables Associated with Death or Transplant at 12 Months of Life

A total of 1,195 patients were 12 months or older at the time of analysis. In total, 24% (288 individuals) either died or were transplanted prior to the first year of life. Survival to 12 months of life was seen in only 57% in the cohort of patients with heart block compared to 79% in those without heart block. (Figure 3a) Heart block was seen more frequently in the death/transplant population than surviving patients (4% vs 1%, p=0.006).(Table 4) In comparison of heart block type, third degree heart block was associated with death or transplant (3% vs. 0.4%, p=0.0007) though second degree heart block was not (2% vs. 1%, p=0.3). Patients with third degree heart block had worse survival compared to those with second degree heart block. (Figure 3b) Heart block patients with a pacing system had slightly lower mortality than those without, though not statistically significant. In addition to heart block, primary diagnosis, the presence of a major syndrome or congenital anomaly, and surgical type were associated with death or transplantation prior to 12 months of age. A total of 10 pacemakers were implanted during stage I admission and 1 implanted in the interstage period, with no difference in associated transplant free 12 month survival (p=0.3). (Figure 3c) On multivariable analysis, heart block (OR 4.9 (1.8–17.5), p=0.01), the presence of an additional non-cardiac syndrome (OR 1.7 (1.1-2.5, p=0.01) and hybrid procedure (OR 2.4 (1.5–3.7), p=0.0001) were associated with increased risk of death while the use of digoxin at Stage I discharge (OR 3.8(2.9-5.1), p=<0.0001) was associated with increased survival. (Table 2)

## Discussion

Perioperative heart block in single ventricle patients is relatively rare but is associated with significant potential morbidity and mortality.(12) Patients with permanent heart block require long term ventricular pacing and pacemaker implantation, which has been associated with long term ventricular dysfunction in older age Fontan cohorts.(7,9) Additionally, in those patients with early recovery of AV nodal function after transient heart block, the recurrence risk of heart block is unknown. In this study, the overall risk of heart block in this large cohort of patients was ~2% and associated with markedly lower 12 month survival particularly in patients with complete heart block as opposed to second degree block. In this diverse population, heterotaxy and atrial arrhythmias were associated with heart block surrounding stage I palliation.

While the exact incidence of heart block in larger single ventricle populations is unclear, in the reported literature it has been seen in 4–11%.(4) In this study of patients from a diverse hospital and regional mix, the incidence of heart block was slightly lower than

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previously reported at  $\sim 2\%$ . Whether this difference is secondary to the larger cohort size and more diverse patient make up is unknown, but is likely to represent a reasonable incidence of heart block in the current era. Interestingly, this data also differed from prior studies with regard to factors associated with heart block around stage I palliation. In recent data from the Pediatric Heart Network, the need for additional AV valve surgery during the stage I admission was found to be associated with heart block, though this association was not seen in the single ventricle sub population.(4–6) Despite a similar incidence in need for additional tricuspid valve surgery, it accounted for only 7% of heart block patients in this cohort as compared to 15% in the previous studies.(4,5) Similarly, while heterotaxy syndrome in non-single ventricle populations has been demonstrated to be associated with a variety of conduction system diseases, smaller studies have not demonstrated a link between heart block and heterotaxy, and larger studies have not evaluated the relationship in this population. This study demonstrated a strong relationship between heterotaxy syndrome and heart block and future studies evaluating the mechanistic drivers may be warranted.(6) The relationship between heart block and atrial arrhythmias is of interest though the cause/effect relationship is unclear. Whether heart block is driving secondary atrial arrhythmias or there is a yet undetermined secondary factor driving the relationship is difficult to determine. While patients with heart block may be more likely to have atrial arrhythmias secondary to resultant hemodynamic embarrassment or possibly secondary to the need for associated pacing, a direct cause and effect cannot be determined from this dataset and requires additional study.

The deleterious effects of heart block and chronic ventricular pacing have been difficult to delineate in the early care of single ventricle populations secondary to small patient cohorts. In several smaller studies, heart block and requirement for pacing has been evaluated in the interstage period but this has not been studied at later time points. In addition to worse survival, heart block was also associated with interstage readmission underscoring the increased complexity of this population throughout their care cycle. Difference between this and prior data may stem from prior studies' significant limitation of using a broad definition of heart block without delineation between second and third degree heart block.(4,9) In this study, early heart block during stage I palliation was found to have increased risk for death out to 12 months of life, but of importance, this risk seemed to be restricted to patients with complete or third degree heart block rather than in patients with second degree block.

Interestingly, only 29% of patients with heart block underwent permanent pacemaker implant. The placement of a permanent pacing system was not statistically significant, but did show a non-significant improved 12 month outcome. This sub-cohort comparison was extremely small and not adequately powered and given pacing is the only current feasible treatment option, this needs further study. Unfortunately, transient heart block with return of normal AV nodal conduction is not specifically captured in the database. What role transient heart block or the recurrence of transient heart block plays in 12 month outcomes remains unknown, though only a single patient in the heart block cohort was readmitted after stage 1 discharge for pacemaker implantation. While it would be assumed that single ventricle patients with complete heart block were not discharged to home without stable pacing, a greater proportion of patients with heart block did remain admitted through the interstage period. What role heart block played in that decision and whether the presence of transient

heart block or persistent second degree heart block was felt to place patients at high risk and led to lack of interstage discharge is unknown and requires additional study.

## Limitations

There are several limitations, primarily related to use of large databases and the rarity of heart block. While allowing for a broad perspective of patients from different centers, analysis is limited to collected data fields. Several variables of interest were unable to be analyzed. One of these variables was the duration and persistence (transient vs. permanent) of 3<sup>rd</sup> degree heart block, particularly in patients who did not receive pacemaker implantation. While we would presume it unlikely for a single ventricle patient to be discharged with complete heart block without stable pacing, the duration of the heart block would be of interest and will require additional evaluation. Due to the low rate of heart block, confidence intervals around the odds ratios remain wide.

#### Conclusions

Single ventricle patients with complete heart block during stage I palliation are a particularly high risk group of patients. The presence of heterotaxy syndrome was the only identified mechanistic driver associated with heart block in this cohort and adds to the complexity of this patient population. Heart block was associated with lower transplant free survival though only in the sub-cohort of patients with third degree heart block as opposed to patients with second degree block. The role of transient heart block in deleterious outcomes and potential rescue with long term pacing remains unknown and requires additional study.

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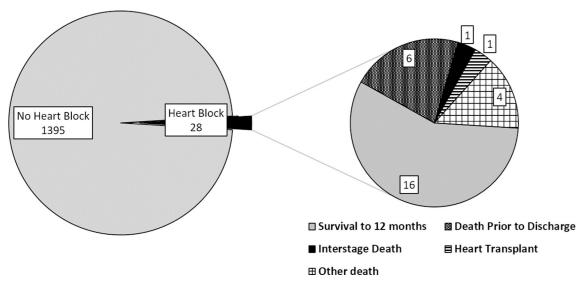
## Abbreviations/Glossary

NPC-QIC	National Pediatric Cardiology Quality Improvement Collaborative
S1P	Stage 1 palliation
HLHS	hypoplastic left heart syndrome
LOS	Length of stay

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## **Outcomes of Patients with Heart Block**

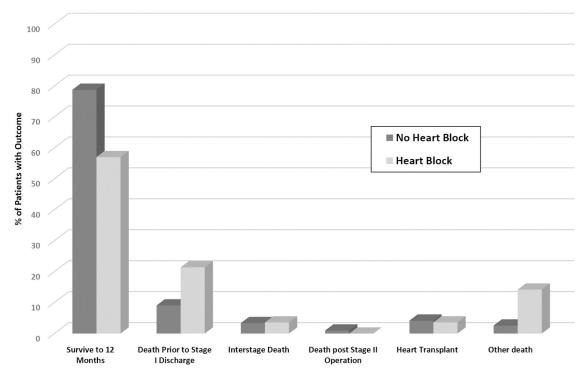


## Figure 1.

Outcomes of Patients with Heart Block

The orange wedge represents the 2% of the total population with heart block at Stage I palliation.

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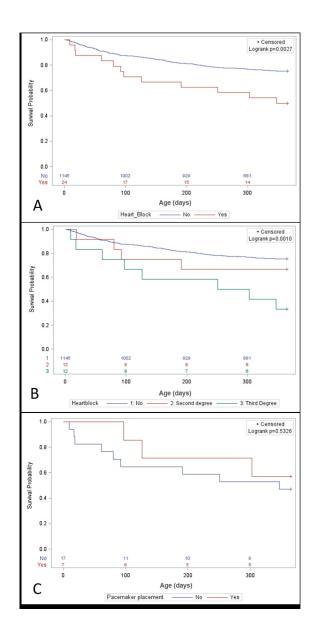


#### Figure 2.

Comparative Outcomes in Patients with and without Heart Block

The blue bar represents patients with no heart block. The orange bar represents patients with heart block during stage I palliation.

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#### Figure 3.

Outcomes of Patients with Heart Block by Heart Block Type and Pacemaker A. Kaplan Meier Curve comparing patients with and without heart block over the first 12 months of life.

**B.** Kaplan Meier Curve comparing patients by degree of heart block.

**C.** Kaplan Meier Curve comparing heart block patients by the presence or absence of a permanent pacing system.

**D.** Kaplan Meier Curve comparing patients with 1) heterotaxy syndrome with no heart block; 2) patients with second degree heart block and 3) patients with third degree heart block.

## Table 1.

Patient and Surgical Characteristics in Patients with and without Heart Block

	Heart Block (n=28)	No Heart Block (n=1395)	P-valu
Stage I Palliation Data, mean±SD			
Age at time of Stage I Palliation (days)	13.7±18	9.6±16	0.08
Weight at Stage I Palliation (kg)	3.2±0.6	3.3±0.6	0.8
Stage II Palliation Data, mean±SD			
Age at time of Stage II Palliation (days)	183.3±67	152.6±45	0.03
Weight at Stage II Palliation (kg)	6.5±1	6.2±1	0.2
Female, n (%)	12 (43%)	554 (40%)	0.9
Race, n (%)			
White	18 (64%)	958 (69%)	
Black-African American	2 (7%)	214 (15%)	
Asian	2 (7%)	8 (1%)	0.05
American Indian	1 (4%)	10 (1%)	
Other	5 (18%)	213 (15%)	
Primary Diagnosis, n (%)			
Hypoplastic Left Heart Syndrome	19 (68%)	1037 (74%)	
Double Inlet Left Ventricle	2 (7%)	69 (5%)	
Double Inlet Right Ventrcle	0 (0%)	4 (0%)	
Mitral Atresia	0 (0%)	29 (2%)	
Tricuspid Atresia	0 (0%)	44 (3%)	0.6
Unbalanced Atrio-ventricular Canal	2 (7%)	76 (5%)	
Doutlet Outlet Right Ventrcle	0 (0%)	12 (1%)	
Single Ventricle Other	1 (4%)	46 (3%)	
Other	4 (14%)	86 (6%)	
Heterotaxy Syndrome, n (%)	3 (11%)	26 (2%)	0.02
Major Congenital Anomaly of Other Organ, n (%)	4 (14%)	92 (7%)	0.2
Type of Surgery, n (%)			
Norwood with BTS	6 (21%)	379 (27%)	
Norwood with RV-PA Conduit	18 (64%)	776 (56%)	
Hybrid Procedure	2 (7%)	106 (8%)	
Norwood Central Shunt	0 (0%)	15 (1%)	0.6
DKS with BT Shunt	0 (0%)	27 (2%)	
DKS with RV-PA Conduit	2 (7%)	50 (4%)	
Other	0 (0%)	42 (3%)	
Additional AV Valve Repair, n (%)	2 (7%)	34 (2%)	0.2

	Heart Block (n=28)	No Heart Block (n=1395)	P-value
Post Operative Rhythm Abnormalities, n (%)			
Atrial Flutter	3 (11%)	44 (3%)	0.03
Chaotic Atrial Rhythm	0 (0%)	9 (1%)	0.6
Ectopic Atrial Tachycardia	3 (11%)	135 (10%)	0.8
Junctional Ectopic Atrial Tachycardia	1 (4%)	66 (5%)	0.8
Re-entrant SVT	3 (11%)	152 (11%)	1
Sinus Bradycardia	0 (0%)	50 (4%)	0.3
Ventricular Fibrillation	0 (0%)	12 (1%)	0.6
Ventricular Tachycardia	3 (11%)	58 (4%)	0.2
Pacemaker Implant Prior to Discharge, n (%)	8 (29%)	3 (0%)	<0.000
Ventricular Function Prior to Stage I Discharge, n (%	o)		
Normal or low normal	11 (39%)	997 (71%)	
Mild Dysfunction	1 (4%)	63 (5%)	
Moderate Dysfunction	0 (0%)	18 (1%)	0.7
Severe Dysfunction	0 (0%)	4 (0%)	
No information	1 (4%)	15 (1%)	
Digoxin at Discharge, n (%)	9 (32%)	755 (54%)	0.02
Death, n (%)	11 (39%)	225 (16%)	0.001

#### Table 2.

#### Multivariable Model of Associated Risks of Heart Block and Death Prior to 12 Months

	Odds Ratio (95%CI)	p-value
Associated Risk for Heart Block During Stage I Admission		
Heterotaxy Syndrome	6.4 (1.8–22.7)	0.004
Atrial Flutter/fibrillation	3.8 (1.1–13.0)	0.04
Associated Risk of Death 1 1101 to 12 Wolfuls		
Heart Block	4.9 (1.4–17.5)	0.01
Associated Risk of Death Prior to 12 Months Heart Block Non-cardiac Syndrome	4.9 (1.4–17.5) 1.7 (1.1–2.5)	0.01 0.01
Heart Block	· · · · ·	

#### Table 3.

#### Outcomes Based on the Presence/Absence of Heart Block

	Heart Block	No Heart Block	P-value
Stage I Palliation Length of Stay	45.2+30	38.2+27	0.1
Age at Discharge Stage I (days)	62.4±40	47±31	0.05
Patient Weight At Discharge Stage I (kg)	3.9±1	3.8±1	0.3
Sage II Palliation Length of Stay	31.0+63	18.7+23	0.4
Remained Inpatient to Stage II Palliation	8 (29%)	152 (11%)	0.003
Unanticipated Interstage Readmission	5 (18%)	485 (35%)	0.4
Admission with PCM Implant	1 (4%)	1 (0%)	< 0.000
Outcomes			
Survive to 12 months	16 (57%)	1100 (79%)	0.002
Heart Transplant	1 (4%)	59 (4%)	0.9
Survival Outcome Comparison			
Survive to 12 months	16 (57%)	1100 (79%)	0.002
Death Prior to Discharge	6 (21%)	127 (9%)	
Interstage Death	1 (4%)	48 (3%)	
Death post Stage II	0 (0%)	14 (1%)	
Heart Transplant	1 (4%)	59 (4%)	
Other death	4 (14%)	36 (3%)	
Cause of Death Arrhythmia, n (%)	0 (0%)	9 (1%)	0.5

#### Table 4.

#### Survival Comparison at 12 Months of Life

Race         White         629 (69%)         193 (67%)           Black-African American         143 (16%)         40 (14%)           Asian         5 (1%)         3 (1%)         0.1           American Indian         5 (1%)         4 (1%)         0.1           Other         125 (14%)         56 (19%)         100           Primary Diagnosis         Hypoplastic Left Heart Syndrome         664 (73%)         227 (79%)           Double Inlet Left Ventricle         49 (5%)         5 (2%)         0.02           Mitral Atresia         18 (2%)         6 (2%)         100%)           Mitral Atresia         13 (3%)         4 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         000           Double Outlet Right Ventricle         2 (0%)         2 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.000           Major Syndrome         88 (10%)         56 (19%)         <0.000           Major Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.000           Type of Surgery         Norwood with BTS         259 (29%)		Survival (n=907)	Death/ Transplant (n=288)	P-valu
White         629 (69%)         193 (67%)           Black-African American         143 (16%)         40 (14%)           Asian         5 (1%)         3 (1%)         0.1           American Indian         5 (1%)         4 (1%)         0           Other         125 (14%)         56 (19%)         1           Primary Diagnosis         Hypoplastic Left Heart Syndrome         664 (73%)         227 (79%)           Double Inlet Left Ventricle         49 (5%)         5 (2%)         0.02           Double Inlet Right Ventricle         2 (2%)         1 (0%)         0.02           Unbalanced Arrioventricular Canal         43 (5%)         24 (8%)         0.02           Unbalanced Arrioventricular Canal         43 (5%)         20 (7%)         0.02           Major Syndrome         88 (10%)         56 (19%)         <0.000           Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         31 (1%)         0.000           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)         Norwood central Shunt         8 (1%)         3 (1%)           Norwood with BTS         259 (29%)         64 (22%)         Norwood central Shunt         2 (2%)	Female, n(%)	344 (38%)	132 (46%)	0.1
Black-African American         143 (16%)         40 (14%)           Asian         5 (1%)         3 (1%)         0.1           American Indian         5 (1%)         4 (1%)         0.1           Other         125 (14%)         56 (19%)         1           Primary Diagnosis         Hypoplastic Left Heart Syndrome         664 (73%)         227 (79%)           Double Inlet Left Ventricle         49 (5%)         5 (2%)         0.02           Mitral Atresia         18 (2%)         6 (2%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outle Right Ventricle         2 (0%)         2 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outle Right Ventricle         2 (0%)         2 (1%)         56 (19%)           Major Syndrome         88 (10%)         56 (19%)         <0.000	Race			
Asian         5 (1%)         3 (1%)         0.1           American Indian         5 (1%)         4 (1%)         0.1           Other         125 (14%)         56 (19%)         1           Primary Diagnosis         Hypoplastic Left Heart Syndrome         664 (73%)         227 (79%)           Double Inlet Left Ventricle         49 (5%)         5 (2%)         0.02           Mitral Atresia         18 (2%)         6 (2%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outle Right Ventricle         2 (0%)         2 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outle Right Ventricle         2 (0%)         2 (1%)         56 (19%)           Single Ventricle Other         35 (39%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0002           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)         0.000           Norwood with RV-PA Conduit         500 (55%)         147 (51%)         0.000           DKS with BT Shunt         22 (2%)         4 (1%)         0.000	White	629 (69%)	193 (67%)	
American Indian         5 (1%)         4 (1%)           Other         125 (14%)         56 (19%)           Primary Diagnosis	Black-African American	143 (16%)	40 (14%)	
Other         125 (14%)         56 (19%)           Primary Diagnosis         Hypoplastic Left Heart Syndrome         664 (73%)         227 (79%)           Double Inlet Left Ventricle         49 (5%)         5 (2%)         Double Inlet Right Ventricle         2 (2%)         1 (0%)           Mitral Atresia         18 (2%)         6 (2%)         Tricuspid Atresia         31 (3%)         4 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         Double Outlet Right Ventricle         2 (0%)         2 (1%)           Single Ventricle Other         35 (39%)         7 (2%)         0.02           Other         63 (7%)         20 (7%)         0           Major Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0002           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)         0.400           Norwood with BTS         259 (29%)         64 (22%)         0.000         0           DKS with BT Shunt         22 (2%)         4 (1%)         0.000         0           DKS with BT Shunt         22 (2%)         4 (1%)         0.002         0.000         0         0.002         0.	Asian	5 (1%)	3 (1%)	0.1
Primary Diagnosis         Hypoplastic Left Heart Syndrome         664 (73%)         227 (79%)           Double Inlet Left Ventricle         49 (5%)         5 (2%)         Double Inlet Right Ventricle         2 (2%)         1 (0%)           Mitral Atresia         18 (2%)         6 (2%)         Tricuspid Atresia         31 (3%)         4 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outlet Right Ventricle         2 (0%)         2 (1%)         Single Ventricle Other         35 (39%)         7 (2%)           Other         63 (7%)         20 (7%)         0.8         40%         0.0002           Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8         0.0002           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)         0.0002           Type of Surgery         Norwood S8 (6%)         47 (16%)         <0.000	American Indian	5 (1%)	4 (1%)	
Hypoplastic Left Heart Syndrome         664 (73%)         227 (79%)           Double Inlet Left Ventricle         49 (5%)         5 (2%)           Double Inlet Right Ventricle         2 (2%)         1 (0%)           Mitral Atresia         18 (2%)         6 (2%)           Tricuspid Atresia         31 (3%)         4 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outlet Right Ventricle         2 (0%)         2 (1%)         Single Ventricle Other         35 (39%)         7 (2%)           Other         63 (7%)         20 (7%)         0.000         410%)         0.0000           Major Syndrome         19 (2%)         7 (2%)         0.8         0.0000           Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0000           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)         0.8           Norwood Central Shunt         8 (1%)         3 (1%)         4 (1%)         0.000           DKS with BT Shunt         22 (2%)         4 (1%)         0.002           DKS with RV-PA Conduit         39 (4%)         9 (3%)	Other	125 (14%)	56 (19%)	
Double Inlet Left Ventricle         49 (5%)         5 (2%)           Double Inlet Right Ventricle         2 (2%)         1 (0%)           Mitral Atresia         18 (2%)         6 (2%)           Tricuspid Atresia         31 (3%)         4 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outlet Right Ventricle         2 (0%)         2 (1%)         5           Single Ventricle Other         35 (39%)         7 (2%)         0.02           Major Syndrome         88 (10%)         56 (19%)         <0.000	Primary Diagnosis			
Double Inlet Right Ventricle         2 (2%)         1 (0%)           Mitral Atresia         18 (2%)         6 (2%)           Tricuspid Atresia         31 (3%)         4 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)         0.02           Double Outlet Right Ventricle         2 (0%)         2 (1%)         Single Ventricle Other         35 (39%)         7 (2%)           Other         63 (7%)         20 (7%)         0.000         Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8           Major Syndrome         19 (2%)         7 (2%)         0.8         0.0002           Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0002           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)         Norwood           Norwood Central Shunt         8 (1%)         3 (1%)         40.000         DKS with BT Shunt         22 (2%)         4 (1%)           DKS with BT Shunt         22 (2%)         4 (1%)         0.02         0.000         0.02           Post Operative Rhythm Abnormalities Heart Block         282 (31%)         130 (45%)         <0.000	Hypoplastic Left Heart Syndrome	664 (73%)	227 (79%)	
Mitral Atresia         18 (2%)         6 (2%)           Tricuspid Atresia         31 (3%)         4 (1%)         0.02           Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)           Double Outlet Right Ventricle         2 (0%)         2 (1%)           Single Ventricle Other         35 (39%)         7 (2%)           Other         63 (7%)         20 (7%)           Major Syndrome         88 (10%)         56 (19%)         <0.000	Double Inlet Left Ventricle	49 (5%)	5 (2%)	
Tricuspid Atresia       31 (3%)       4 (1%)       0.02         Unbalanced Atrioventricular Canal       43 (5%)       24 (8%)         Double Outlet Right Ventricle       2 (0%)       2 (1%)         Single Ventricle Other       35 (39%)       7 (2%)         Other       63 (7%)       20 (7%)         Major Syndrome       88 (10%)       56 (19%)       <0.000	Double Inlet Right Ventricle	2 (2%)	1 (0%)	
Unbalanced Atrioventricular Canal         43 (5%)         24 (8%)           Double Outlet Right Ventricle         2 (0%)         2 (1%)           Single Ventricle Other         35 (39%)         7 (2%)           Other         63 (7%)         20 (7%)           Major Syndrome         88 (10%)         56 (19%)         <0.000	Mitral Atresia	18 (2%)	6 (2%)	
Double Outlet Right Ventricle         2 (0%)         2 (1%)           Single Ventricle Other         35 (39%)         7 (2%)           Other         63 (7%)         20 (7%)           Major Syndrome         88 (10%)         56 (19%)         <0.000           Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0002           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)           Norwood with RV-PA Conduit         500 (55%)         147 (51%)           Norwood Central Shunt         8 (1%)         3 (1%)           Hybrid Norwood         58 (6%)         47 (16%)         <0.000	Tricuspid Atresia	31 (3%)	4 (1%)	0.02
Single Ventricle Other         35 (39%)         7 (2%)           Other         63 (7%)         20 (7%)           Major Syndrome         88 (10%)         56 (19%)         <0.000	Unbalanced Atrioventricular Canal	43 (5%)	24 (8%)	
Other         63 (7%)         20 (7%)           Major Syndrome         88 (10%)         56 (19%)         <0.000	Double Outlet Right Ventricle	2 (0%)	2 (1%)	
Major Syndrome         88 (10%)         56 (19%)         <0.000           Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0007           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)           Norwood with RV-PA Conduit         500 (55%)         147 (51%)           Norwood Central Shunt         8 (1%)         3 (1%)           Hybrid Norwood         58 (6%)         47 (16%)         <0.000	Single Ventricle Other	35 (39%)	7 (2%)	
Heterotaxy Syndrome         19 (2%)         7 (2%)         0.8           Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0002           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)           Norwood with RV-PA Conduit         500 (55%)         147 (51%)           Norwood Central Shunt         8 (1%)         3 (1%)           Hybrid Norwood         58 (6%)         47 (16%)         <0.000	Other	63 (7%)	20 (7%)	
Major Congenital Anomaly of Other Organ         51 (6%)         36 (13%)         0.0002           Type of Surgery         Norwood with BTS         259 (29%)         64 (22%)           Norwood with RV-PA Conduit         500 (55%)         147 (51%)           Norwood Central Shunt         8 (1%)         3 (1%)           Hybrid Norwood         58 (6%)         47 (16%)         <0.000	Major Syndrome	88 (10%)	56 (19%)	<0.000
Type of Surgery         Norwood with BTS       259 (29%)       64 (22%)         Norwood with RV-PA Conduit       500 (55%)       147 (51%)         Norwood Central Shunt       8 (1%)       3 (1%)         Hybrid Norwood       58 (6%)       47 (16%)       <0.000	Heterotaxy Syndrome	19 (2%)	7 (2%)	0.8
Norwood with BTS         259 (29%)         64 (22%)           Norwood with RV-PA Conduit         500 (55%)         147 (51%)           Norwood Central Shunt         8 (1%)         3 (1%)           Hybrid Norwood         58 (6%)         47 (16%)         <0.000	Major Congenital Anomaly of Other Organ	51 (6%)	36 (13%)	0.0002
Norwood with RV-PA Conduit         500 (55%)         147 (51%)           Norwood Central Shunt         8 (1%)         3 (1%)           Hybrid Norwood         58 (6%)         47 (16%)         <0.000	Type of Surgery			
Norwood Central Shunt         8 (1%)         3 (1%)           Hybrid Norwood         58 (6%)         47 (16%)         <0.000	Norwood with BTS	259 (29%)	64 (22%)	
Hybrid Norwood       58 (6%)       47 (16%)       <0.000	Norwood with RV-PA Conduit	500 (55%)	147 (51%)	
DKS with BT Shunt       22 (2%)       4 (1%)         DKS with RV-PA Conduit       39 (4%)       9 (3%)         Other       21 (3%)       14 (5%)         Additional AV Valve Repair (Y/N)       18 (2%)       13 (1%)       0.02         Post Operative Rhythm Abnormalities Heart Block       282 (31%)       130 (45%)       <0.000	Norwood Central Shunt	8 (1%)	3 (1%)	
DKS with RV-PA Conduit         39 (4%)         9 (3%)           Other         21 (3%)         14 (5%)           Additional AV Valve Repair (Y/N)         18 (2%)         13 (1%)         0.02           Post Operative Rhythm Abnormalities Heart Block         282 (31%)         130 (45%)         <0.000	Hybrid Norwood	58 (6%)	47 (16%)	< 0.000
Other         21 (3%)         14 (5%)           Additional AV Valve Repair (Y/N)         18 (2%)         13 (1%)         0.02           Post Operative Rhythm Abnormalities Heart Block         282 (31%)         130 (45%)         <0.000	DKS with BT Shunt	22 (2%)	4 (1%)	
Additional AV Valve Repair (Y/N)       18 (2%)       13 (1%)       0.02         Post Operative Rhythm Abnormalities Heart Block       282 (31%)       130 (45%)       <0.000	DKS with RV-PA Conduit	39 (4%)	9 (3%)	
Post Operative Rhythm Abnormalities Heart Block         282 (31%)         130 (45%)         <0.000           Any Heart Block         13 (1%)         12 (4%)         0.006           2nd degree         9 (1%)         5 (2%)         0.3           3rd degree         4 (0.4%)         8 (3%)         0.0007	Other	21 (3%)	14 (5%)	
Any Heart Block       13 (1%)       12 (4%)       0.006         2nd degree       9 (1%)       5 (2%)       0.3         3rd degree       4 (0.4%)       8 (3%)       0.0007	Additional AV Valve Repair (Y/N)	18 (2%)	13 (1%)	0.02
2nd degree       9 (1%)       5 (2%)       0.3         3rd degree       4 (0.4%)       8 (3%)       0.0007	Post Operative Rhythm Abnormalities Heart Block	282 (31%)	130 (45%)	< 0.000
3rd degree 4 (0.4%) 8 (3%) 0.0007	Any Heart Block	13 (1%)	12 (4%)	0.006
	2nd degree	9 (1%)	5 (2%)	0.3
Pacemaker Implant         6 (1%)         4 (1%)         0.3	3rd degree	4 (0.4%)	8 (3%)	0.0007
	Pacemaker Implant	6 (1%)	4 (1%)	0.3

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-	Survival (n=907)	Death/ Transplant (n=288)	P-value
Ventricular Function Prior to Stage I Discharge			
Normal or low normal	729 (80%)	99 (34%)	
Mild Dysfunction	50 (6%)	10 (3%)	
Moderate Dysfunction	18 (2%)	2 (1%)	0.3
Severe Dysfunction	2 (0%)	1 (0%)	
No information	16 (2%)	0 (0%)	
Digoxin at Discharge	550 (61%)	79 (27%)	< 0.000
Readmission after Stage I Discharge for PCM	0	1	0.1