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## Neurodevelopmental Outcomes in Preschool Survivors of the Fontan Procedure

J. William Gaynor, M.D.<sup>1</sup>, Richard F Ittenbach, Ph.D.<sup>2</sup>, Marsha Gerdes, Ph.D.<sup>3</sup>, Judy Bernbaum, M.D.<sup>4</sup>, Robert R. Clancy, M.D.<sup>5</sup>, Donna M. McDonald-McGinn, M.S.<sup>6</sup>, Elaine H. Zackai, M.D.<sup>6</sup>, Gil Wernovsky, M.D.<sup>7</sup>, Susan C. Nicolson, M.D.<sup>8</sup>, and Thomas L. Spray, M.D.<sup>1</sup>

<sup>1</sup>Cardiac Surgery, CHOP, Philadelphia, PA, USA

<sup>2</sup>Biostatistics, Cincinnati Children's Hospital, Cincinnati, OH, USA

<sup>3</sup>Psychology, CHOP, Philadelphia, PA, USA

<sup>4</sup>Pediatrics, CHOP, Philadelphia, PA, USA

<sup>5</sup>Neurology, CHOP, Philadelphia, PA, USA

<sup>6</sup>Genetics, CHOP, Philadelphia, PA, USA

<sup>7</sup>Cardiology, CHOP, Philadelphia, PA, USA

<sup>8</sup>Cardiac Anesthesiology

### Abstract

**Objective**—To compare neurodevelopmental (ND) outcomes of preschool survivors of the Fontan procedure with those of children with congenital heart disease (CHD) undergoing biventricular (BV) repair and to investigate predictors of ND outcome for those with single ventricle (SV) CHD, including hypoplastic left heart syndrome (HLHS).

**Methods**—ND outcomes were assessed at 4 years of age including cognition, visual-motor integration, behavior, social skills and academic achievement. Unadjusted outcomes were compared between the BV and SV patients. Predictors of ND outcome were assessed in the SV patients. Multiple-covariate models were evaluated using patient-related, operative, and post-operative covariates.

**Results**—ND evaluation was performed in 365 children, 112 after the Fontan [HLHS ( $n = 91$ ), other SV ( $n = 21$ )] and 253 after BV repair. Compared to BV patients, the SV patients performed worse in terms of processing speed, inattention and impulsivity. Otherwise there were no significant differences between the groups for any domain. There was a trend toward lower performance for SV patients on visual motor integration. Outcomes for patients with HLHS were

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Corresponding Author: J. William Gaynor, M.D., Division of Cardiothoracic Surgery, The Children's Hospital of Philadelphia, 34<sup>th</sup> and Civic Center Boulevard, Philadelphia, PA 19104, USA, Phone: 215-590-2708 / Fax: 215-590-2715, gaynor@email.chop.edu. From The Cardiac Center, The Children's Hospital of Philadelphia, Philadelphia, PA, USA

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not worse than for other forms of functional SV. Patient factors were more important predictors of ND outcomes than were operative management variables.

**Conclusions**—In this cohort, unadjusted ND outcomes for preschool survivors of the Fontan procedure are similar to those for children with CHD undergoing BV repair for most domains. Among the Fontan patients, HLHS was not associated with worse outcomes compared to other forms of SV.

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## Introduction

Over the past 15 years, innovations in surgical techniques and perioperative care have resulted in improving survival for patients with the most complex forms of congenital heart disease (CHD); especially those born with functional single ventricle (SV), including hypoplastic left heart syndrome (HLHS).<sup>1</sup> With improved survival has come the realization of neurobehavioral disabilities and impaired functional outcomes in a significant portion of the survivors. Indeed, for all children with CHD, neurodevelopmental dysfunction has become the most common and potentially the most disabling outcome for CHDs and their treatment.

Patients with SV who ultimately undergo the Fontan operation are at the greatest risk for neurodevelopmental (ND) disability. These patients usually undergo multiple surgical procedures with cardiopulmonary bypass (CPB), and often deep hypothermic circulatory arrest (DHCA). Hospitalizations are multiple, and are typically longer than for those with a biventricular (BV) circulation. In addition, before the completion of the Fontan operation all patients with SV have chronic hypoxemia, which may be a significant risk factor for later cognitive dysfunction and ND abilities. Previous studies have suggested that as a group, patients with SV function within the low normal range for cognitive performance and many other developmental domains.<sup>2-4</sup>

The current study was undertaken to assess ND performance in multiple domains for children with various forms of SV undergoing staged reconstruction surgery and ultimately the Fontan procedure, as well as to 1) compare the ND outcomes to children with other forms of CHD who underwent a BV repair, and 2) evaluate potential risk factors for adverse ND outcomes in the cohort of children with SV.

## Materials and Methods

The current study is a secondary analysis of data from a prospective longitudinal study evaluating the association between neurodevelopmental dysfunction and polymorphisms of the apolipoprotein E (*APOE*) gene in preschool patients (4–5 years of age) after cardiac surgery.<sup>5</sup> Patients 6 months of age or younger undergoing surgery for CHD using CPB with or without DHCA were eligible. Exclusion criteria included multiple congenital anomalies, a recognizable genetic syndrome other than chromosome 22q11 microdeletion syndrome and language other than English spoken in the home. The Institutional Review Board at The Children's Hospital of Philadelphia approved the study and the parent or guardian provided informed consent.

The population for the current study consisted of all patients who had returned for neurodevelopmental evaluation at 4 to 5 years of age and who had achieved an end-state of either a biventricular repair or a Fontan procedure. Children who had not achieved one of these end-states or who had undergone cardiac transplantation were not included in the analysis.

### **Operative Management**

Operations were performed by five cardiac surgeons with a dedicated team of cardiac anesthesiologists. Alpha-stat blood gas management was used. Pump flow rates were not standardized for this study. DHCA was used at the surgeon's discretion. Before DHCA, patients underwent core cooling and topical hypothermia of the head to a nasopharyngeal temperature of 18°C. Modified ultrafiltration was performed in all patients. Patients recovered in a Cardiac Intensive Care Unit with a dedicated group of cardiac intensivists.

### **Data Collection**

Preoperative factors including gestational age, birth head circumference, birth weight, and preoperative intubation were obtained from birth and hospital records. Weight, age at operation, and type of operation were recorded along with perfusion data, including CPB time, aortic cross-clamp time, and duration of DHCA. Total support time was calculated as CPB time plus DHCA time. Total DHCA time was calculated as the sum of the duration of each episode of DHCA.

### **Four-year Neurodevelopmental Examination**

The neurodevelopmental examination was performed between the 4<sup>th</sup> and 5<sup>th</sup> birthdays; Maternal education, socioeconomic status, and ethnicity were determined by parental report. A medical history was obtained focusing on illness, rehospitalizations, neurologic events or interim evaluations, current medications, and parent concerns over health.

Patients were evaluated by a genetic dysmorphologist at 1 and/or 4 year evaluations. Additional genetic analyses were performed if indicated. Neonatal recognition of dysmorphic features can be difficult; therefore, some patients were enrolled for whom the diagnosis of a genetic syndrome was not made until a later evaluation. Patients were classified as having no definite syndromic/chromosomal abnormality (normal), a suspected genetic syndrome (suspect), or definite syndromic/chromosomal abnormality (abnormal). APOE genotype determination, whole blood, or a buccal swab was obtained before the operation and stored at 4° centigrade. Genomic DNA was prepared to determine the APOE genotype using a previously published method.<sup>5</sup>

Cognitive outcomes were assessed using the Wechsler Preschool and Primary Scale of Intelligence, Third Edition (WPPSI-III).<sup>6</sup> The WPPSI-III provides 4 scales: Verbal IQ (VIQ) estimates verbal reasoning and comprehension and attention to verbal stimuli. Performance IQ (PIQ) estimates non-verbal reasoning including fluid reasoning, spatial processing and perceptual-organization. Full scale IQ (FSIQ) is a summary of both types of reasoning. Processing speed (PS) estimates the ability to process information without making errors. Visual-motor integration was assessed with the Developmental Test of Visual Motor

Integration (VMI), a simple copying task that assesses the child's fine-motor and visual-motor coordination skills.<sup>7</sup> Academic achievement (school readiness for reading and math) was tested using the reading and math clusters of the Woodcock-Johnson III, a standardized achievement test for children from 2 year to adulthood.<sup>8</sup> If a child was judged to be too developmentally impaired to complete the tasks, he/she was assigned the lowest possible score for the specific test; if a child was unable to complete the task for other reasons, the child was excluded from the analysis for that domain.

Inattention, impulsivity, and social skills were assessed by parental report. Inattention and impulsivity were also assessed by the Impulsivity and Inattention Scales of the Attention Deficit/Hyperactivity Disorder (ADHD) Rating Scale-IV Preschool Version.<sup>9</sup> Social Competence was assessed by the Preschool and Kindergarten Behavior Rating Scales (PKBS) Social Skills Total Score which details social cooperation, social interaction and social independence as reported by parents.<sup>10</sup> Social interaction skills were also assessed using the Child Behavior Checklist for ages 1.5 to 5 years (CBCL/1.5–5).<sup>11</sup> The CBCL/1.5–5 is a questionnaire used to obtain parental reports of behavior problems and prosocial adaptive skills demonstrated within the previous 6 months. Specifically, the pervasive developmental problem (PDP) scale was utilized to assess the prevalence of problems in the area of reciprocal social interactions and restricted behaviors (e.g., repetitive behavior or disturbed by change). The PDP scale was developed to incorporate some of the behavioral symptoms that the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) lists as criteria for the diagnosis of an autism spectrum disorder (autism, Asperger syndrome, or pervasive developmental disorder not otherwise specified). High scores on the PDP scale do not confirm the diagnosis of an autism spectrum disorder but suggest that further evaluation is warranted.

## Data Analysis

Data analysis proceeded in three distinct phases, a descriptive phase in which we computed and evaluated the descriptive information for the group as a whole and by subgroup (Fontan, BV), a comparison phase in which we compared the distributions of standardized scores of each neurodevelopmental outcome between the BV and SV patients, and, finally, a modeling phase in which we identified risk factors for each of the aforementioned ND outcomes in the SV patients. A more complete description of each phase is provided below. All data were analyzed using SAS v9.3.

**Descriptive Phase**—Simple descriptive statistics were computed using both parametric and nonparametric measures of central tendency, variability and association for all relevant variables in the data set. Measures of skewness and kurtosis were evaluated for all outcomes to test for normality and subsequent modeling. Histograms and frequency counts were generated for variables that were dichotomous or categorical in nature.

**Comparison Phase**—Two sets of comparisons were computed; the first involved comparing children with a Fontan circulation with children with BV circulation across a series of patient-related and surgical variables. The two groups were then compared with respect to level of impairment for cognitive function, visual-motor integration, academic

achievement and social skills. All of these instruments have an expected mean of 100, with a standard deviation (SD) of 15. Moderate impairment for all tests was defined as a score between 1 SD and 2 SD less than the expected mean. Severe impairment was defined as a score more extreme than 2 SD below the expected mean. In the normative population, approximately 16% will have scores more extreme than 1 SD below the mean and approximately 2.5% will have scores more extreme than 2 SDs below the mean. Inattention, impulsivity, social skills and restricted behaviors were also compared between groups. For both sets of comparisons, Wilcoxon rank-sum tests were used for data that were continuous in nature, including ordered categorical data, and Chi-square tests of association were used for data that were categorical nature. Fisher's Exact tests were used when expected cell counts were less than five.

**Modeling Phase**—In the risk-factor modeling phase, a series of 20 single covariate generalized linear models were specified and tested for each of the 11 aforementioned outcomes. The four cognitive scores, the visual motor scores, and the two academic achievement scores were all evaluated using a generalized linear model with a Gaussian (normal) distribution and an identity link, while the four behavioral and social skills scores were evaluated using models with a Poisson distribution and a log link. Because of the rather skewed nature of the achievement scores, both math achievement and reading achievement scores were transformed as follows: WJ Math <sup>(1.96)</sup> and reading achievement <sup>(-0.55)</sup> using Box-Cox transformations. Multiple covariate models were then evaluated using candidate covariates with Wald statistic *p*-values < 0.05. The criterion for statistical significance for all models, single- as well as multiple-covariate, was set at the unadjusted  $\alpha = 0.05$  level.

## Results

### Study Group

Between September 1998 and April 2003, 675 eligible infants underwent cardiac surgery. A total of 23 infants died before consent was obtained while parents of 102 infants declined participation in the study, resulting in 550 infants (81%) enrolled. There were 21 deaths during the initial hospitalization and an additional 43 deaths prior to 5 years of age. A total of 486 patients were eligible for the 4 year evaluation which was completed by 381 patients (78% of all eligible patients). Baseline characteristics were compared for patients who returned for the 4 year evaluations, those who did not return ( $n = 105$ ), and those who died before 4 years of age ( $n = 64$ ). The only statistically significant difference in baseline characteristics between returning and non-returning patients was under-representation of black patients among the returning patients (21% versus 29%).<sup>12</sup>

Patients were considered for enrollment in the current analysis if they had achieved an end-state of either a completed BV repair or a Fontan operation. Patients who had not achieved one of these end-states or who had undergone cardiac transplantation were not considered in this analysis. A total of 365 patients met entry criteria for this analysis. BV repair was been achieved in 253 patients and 112 patients had undergone the Fontan procedure. Preoperative and demographic variables are listed in table 1. There were no significant differences between the biventricular repair patients and the SV patients in terms of gender, ethnicity,

gestational age, birth weight, birth head circumference, maternal education or socioeconomic status. Presence of a definite genetic anomaly was more common in patients undergoing biventricular repair (17%) compared to the patients undergoing the Fontan procedure (6%),  $p < 0.01$ . Patients who underwent the Fontan operation were more likely to have mechanical ventilation prior to the initial operation,  $p < 0.01$ .

Operative management variables are shown in table 2 for the initial operation and subsequent operations with CPB prior the ND evaluation. Patients undergoing the Fontan procedure were younger and weighed less at the time of the initial surgery and were more likely to have undergone use of DHCA. Postoperative length of stay (LOS) was also significantly longer for patients who underwent the Fontan procedure. In addition, all patients who underwent the Fontan operation had additional operations with CPB compared to only 11% of patients with biventricular repair group. Almost all patients who underwent the Fontan procedure were also exposed to additional periods of DHCA.

### **Neurodevelopmental Outcomes: Biventricular Repair vs. Fontan Procedure**

Unadjusted outcomes for all the neurodevelopmental domains tested are shown in table 3 for the entire cohort as well as for the BV repair patients and the Fontan patients. Scores for the entire cohort were generally in the low normal range for all domains tested. Comparison of the biventricular repair patients and Fontan patients show that ND outcomes are similar. The Fontan patients performed significantly worse in terms of processing speed and demonstrated significantly higher scores (worse performance) for inattention and impulsivity. Otherwise there were no statistically significant differences between the BV repair patients and the SV patients for any domain tested. There was a trend towards lower performance among the SV patients on visual motor integration ( $p = 0.06$ ).

The prevalence and severity of moderate and severe impairments were also compared between the BV repair patients and the SV patients (table 4). For most domains evaluated, the number of patients with moderate to severe impairment was greater than expected for the general population, but the majority of children (74 to 92%) performed within the normal range. The pattern of impairment was not different between the BV repair patients and the Fontan patients for most domains. In the Fontan group, there were more patients with moderate impairment of visual-motor integration,  $p = 0.02$ . There was a trend towards greater impairment of processing speed, a measure of executive function, in the Fontan patients ( $p = 0.09$ ).

### **Predictors of Neurodevelopmental Outcomes after the Fontan Procedure**

With respect to all models, patient factors were more important predictors than were operative management variables. When considering multiple covariate models, in particular, larger birth weight and older gestational age were independently associated with better performance across multiple domains. Presence of a genetic anomaly was an independent predictor of restricted behavior, inattention and impulsivity. Greater Maternal education was associated with better academic achievement. As in previous studies, longer post-operative LOS was associated with worse performance in multiple domains. Importantly, HLHS was not predictive of worse outcomes compared to other forms of functional SV. The results of

the single and multiple covariate models for are shown in tables E1 a–c, available as an electronic appendix..

## Discussion

The results of this study demonstrate, consistent with previous studies, the great majority of preschool survivors of the Fontan procedure perform within the low-normal range for most neurodevelopmental domains. In particular, their performance is equivalent to those patients with a variety of cardiac defects who have undergone BV repair for most domains tested. There has been a general perception that early ND outcomes for patients with functional SV are significantly worse than for children with other types of CHD. The current study suggests that this is no longer true for many ND domains. In addition, HLHS was not a risk factor for worse ND disability compared to other forms of functional SV. Similar to some previous studies, we identified impaired visual-motor integration as a problem for the Fontan patients. We did not find duration of DHCA to be associated with any outcome.

Other than the type of CHD, the patients undergoing biventricular repair and those undergoing the Fontan procedure were similar in terms of patient characteristics. There were no differences with respect to gender, ethnicity, degree of prematurity, birth weight, or birth head circumference. Presence of a definite genetic syndrome was more common in patients undergoing BV repair (17% compared to 6% in the SV patients), and the SV patients were more likely to have required mechanical ventilation prior to the initial intervention. In terms of operative management there were significant differences between the 2 groups. The patients with SV were younger and weighed less at the time of primary surgery. They were more likely to have had use of DHCA and had a longer postoperative length of stay. In addition, all of the patients with functional SV underwent additional operations with cardiopulmonary bypass and most were exposed to additional DHCA. These findings suggest that overall the patients with the Fontan procedures tend to have multiple risk factors for adverse ND outcomes. However, despite the increased risk, ND outcomes in patients with SV are similar to those patients who underwent BV repair.

Several previous studies have evaluated ND outcomes following the Fontan procedure. These often include not just preschool age patients but also older patients. In 1998, Kern and associates evaluated neurodevelopmental outcomes in a small group ( $n = 15$ ) of patients with HLHS at a mean age of 4.4 years.<sup>2</sup> Median scores for full scale IQ and behavior were in the low to normal range. Use of DHCA during the Norwood Procedure was identified as a risk factor for lower IQ. Also, in 1998, Uzark and colleagues from San Diego evaluated 32 survivors of the Fontan procedure.<sup>3</sup> The majority of children functioned within the normal range but below average scores were found for visual-motor skills in 20% of the patients. They found that there was no significant correlation between degree of preoperative hypoxemia prior to the Fontan and ND performance. However, they did identify DHCA during the Norwood procedure as a risk factor for lower IQ. In 2000, Goldberg and colleagues at the University of Michigan evaluated ND outcomes in a cohort of patients with SV.<sup>4</sup> For the group as a whole, ND outcomes were good, with cognitive function generally in the normal range. However, they identified that the patients with HLHS had significantly lower scores than the patients with other forms of functional single ventricle. Although

neither subgroup scored significantly lower than expected population means. They identified lower socioeconomic status, use of DHCA, and perioperative seizures as the predictors of worse ND outcome.

More recently, Sarajuuri and colleagues from Finland examined 27 children with HLHS or other forms of SV at a median of 5.7 years.<sup>13</sup> They found that mean full-scale IQ was significantly lower than the expected population means for both patients with hypoplastic left heart syndrome and for those with other forms of SV. Cerebral palsy was identified in 1 of 7 patients with HLHS and 2 of 10 with other forms of SV. They also performed brain imagining with computed tomography or magnetic resonance imaging and found that abnormal findings correlated significantly with lower full scale IQ and lower verbal IQ.

Brosig and colleagues compared outcomes for preschool children with HLHS and transposition of the great arteries (TGA).<sup>14</sup> Cognitive function and academic function were similar between the groups. However, problems with visual-motor skills and behavior were more common in HLHS patients. We previously compared outcomes for preschool children with Infants with ventricular septal defect (VSD), tetralogy of Fallot (TOF), TGA and HLHS, and no recognized genetic anomalies.<sup>15</sup> Mean scores for the ND domains tested were in the normal range. Unadjusted neurodevelopmental outcomes for HLHS were lower for cognition, fine motor skills, executive function, and math skills compared with the other patients. However after correction for the demographic, preoperative, and operative variables, no significant differences were found among the groups for any domain.

The present study has several limitations. The management of CPB was not standardized for the present study. By the nature of their defect, children with SV experience prolonged exposure to hypoxemia compared with children with BV CHD. However, we did not assess the severity or duration of the hypoxemia. SV patients also had more procedures and more days in hospital than BV patients. Although we evaluated multiple ND domains, these might have provided an incomplete assessment of the potential ND disabilities, especially behavioral problems. In particular, the young age of the children makes an assessment of academic abilities difficult. Finally, for the entire cohort, there was a significant subset of patients who were alive but did not return for the 4-year evaluation. The only difference in baseline and operative characteristics between these patients and those who returned was an under-representation of African-Americans in the returning patients. This difference is unlikely to affect the results of the evaluations significantly.

In conclusion, despite the need for multiple cardiac surgical procedures, unadjusted ND outcomes for preschool aged survivors of the Fontan procedure are similar to those for children with CHD undergoing BV repair for most domains. In contrast to previous reports, among the Fontan patients, HLHS was not associated with worse outcomes compared to other forms of SV. Most children function within the low normal range for the domains tested. The occurrence of severe impairment is greater compared to the general population, but is present in < 10% of patients for most domains tested.



## Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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**Table 1**

## Preoperative and Demographic Variables

Variable	All Patients (n = 365)	Biventricular Repair (n = 253)	Fontan (n = 112)	Biventricular vs. Fontan p-value
Gender				
Male	209 (57%)	141 (56%)	68 (61%)	0.37
Female	156 (43%)	112 (44%)	44 (39%)	
Ethnicity				0.12
White	249 (68%)	167 (66%)	82 (73%)	
Black	75 (21%)	52 (21%)	23 (21%)	
Other	41 (11%)	34 (13%)	7 (6%)	
Gestational Age (weeks)	38.5 ± 2.2	38.5 ± 2.2	38.4 ± 1.9	0.33
Birth Weight (grams)	3,124 ± 637	3,090 ± 639	3,199 ± 630	0.14
Birth Head Circumference (centimeters)	33.6 ± 2.1	33.5 ± 2.0	33.8 ± 2.2	0.06
<b>Genetic Anomaly</b>				<b>&lt; 0.01</b>
<b>Normal</b>	<b>286 (78%)</b>	<b>194 (77%)</b>	<b>92 (82%)</b>	
<b>Suspect</b>	<b>28 (8%)</b>	<b>15 (6%)</b>	<b>13 (12%)</b>	
<b>Abnormal</b>	<b>51 (14%)</b>	<b>44 (17%)</b>	<b>7 (6%)</b>	
Maternal Education				
Less Than High School	19 (5%)	12 (5%)	7 (6%)	0.95
High School/Some College	152 (42%)	106 (42%)	46 (41%)	
College	129 (36%)	90 (36%)	39 (35%)	
Graduate	63 (17%)	44 (17%)	19 (17%)	
Socioeconomic Status				0.81
Professional/medium business	40 (11%)	30 (12%)	10 (9%)	
skilled/clerical	75 (21%)	50 (20%)	25 (22%)	
semi-skilled	115 (31%)	79 (31%)	36 (33%)	
unskilled	133 (37%)	94 (37%)	39 (36%)	
<b>Preoperative Mechanical Ventilation (yes)</b>	<b>110 (30%)</b>	<b>63 (25%)</b>	<b>47 (42%)</b>	<b>&lt; 0.01</b>

**Table 2**

## Operative Variables

Variable	All Patients (n = 365)	Biventricular Repair (n = 253)	Fontan (n = 112)	Biventricular vs. Fontan p-value
<i>First Operation</i>				
Age (days)	43.7 ± 54.4	55.4 ± 56.7	17.1 ± 37.2	< 0.01
Weight (kilograms)	3.9 ± 1.3	4.1 ± 1.3	3.5 ± 1.0	< 0.01
CPB Time (min)	65.6 ± 39.3	70.9 ± 42.9	53.6 ± 25.8	< 0.01
DHCA Used (Yes)	211 (58%)	112 (44%)	99 (88%)	< 0.01
DHCA Time (min)	39.0 ± 16.4	35.8 ± 16.4	42.5 ± 15.8	< 0.01
Hematocrit after Hemodilution (%)	27.9 ± 4.1	27.4 ± 4.1	28.8 ± 3.9	0.01
Postoperative Length of Stay	11.8 ± 12.7	10.7 ± 13.2	14.4 ± 11.3	< 0.01
<i>Subsequent Operations</i>				
Additional Operations with CPB (Yes)	140 (38%)	28 (11%)	112 (100%)	< 0.01
Mean Additional Operations with CPB	0.7 ± 0.9	0.1 ± 0.4	1.9 ± 0.3	< 0.01
Additional CPB Time (min)	100.4 ± 62.5	68.6 ± 50.7	108.4 ± 62.8	0.02
Additional DHCA (Yes)	105 (29%)	7 (3%)	98 (88%)	< 0.01
Additional DHCA Time (min)	45.1 ± 22.6	22.3 ± 13.3	46.7 ± 22.3	< 0.01

**Table 3**

## Unadjusted Neurodevelopmental Outcomes

	All Patients ( <i>n</i> = 365)	Biventricular Repair ( <i>n</i> = 253)	Fontan ( <i>n</i> = 112)	Biventricular vs. Fontan <i>p</i> -value
<b>Cognition</b>				
Full Scale IQ *	95.3 ± 19.0	96.2 ± 19.8	93.3 ± 17.1	0.18
Verbal IQ *	96.6 ± 18.9	96.8 ± 19.6	96.3 ± 17.5	0.80
Performance IQ *	94.9 ± 18.4	95.9 ± 19.1	92.8 ± 16.4	0.14
<b>Processing Speed *</b>	<b>94.7 ± 16.6</b>	<b>96.5 ± 16.9</b>	<b>90.8 ± 15.2</b>	<b>&lt; 0.01</b>
<b>Visual-Motor Integration</b>				
VMI Composite *	92.7 ± 18.2	93.9 ± 18.5	90.0 ± 17.2	0.06
<b>Behavior</b>				
<b>Inattention **</b>	<b>6.3 ± 5.5</b>	<b>6.1 ± 5.3</b>	<b>6.9 ± 5.8</b>	<b>0.01</b>
<b>Impulsivity **</b>	<b>7.2 ± 5.6</b>	<b>6.9 ± 5.5</b>	<b>7.8 ± 5.8</b>	<b>&lt; 0.01</b>
<b>Social Skills</b>				
PKBS Social Skills *	106.2 ± 13.0	106.6 ± 13.4	105.3 ± 12.0	0.28
CBCL Pervasive Developmental Problems **	3.4 ± 3.3	3.3 ± 3.1	3.6 ± 3.8	0.15
<b>Academic Achievement</b>				
Math Achievement *	96.0 ± 20.6	96.9 ± 21.3	94.2 ± 18.7	0.11
Reading Achievement *	106.0 ± 16.8	106.4 ± 16.9	105.1 ± 16.4	0.40

\* Higher score = better performance;

\*\* Higher score = worse performance

**Table 4**

Prevalence and Severity of Impairment

	Biventricular Repair (n = 253)				Fontan (n = 112)				Biventricular vs. Fontan p-value	
	Score 85	Score 71 to 84	Score 70	Score 85	Score 71 to 84	Score 70	Score 85	Score 70		
<b>Cognition</b>										
Full Scale IQ	185 (74%)	45 (18%)	21 (8%)	81 (73%)	20 (18%)	10 (9%)				0.61
Verbal IQ	193 (77%)	38 (15%)	21 (8%)	90 (80%)	14 (14%)	7 (6%)				0.69
Performance IQ	185 (74%)	45 (18%)	21 (8%)	74 (67%)	29 (27%)	7 (6%)				0.17
Processing Speed	162 (72%)	57 (25%)	7 (3%)	61 (61%)	38 (37%)	3 (3%)				0.09
<b>Visual-Motor Integration</b>										
VMI Composite	<b>189 (75%)</b>	<b>36 (14%)</b>	<b>26 (11%)</b>	<b>69 (62%)</b>	<b>29 (26%)</b>	<b>14 (12%)</b>				<b>0.02</b>
<b>Academic Achievement</b>										
Math Achievement	181 (75%)	34 (14%)	25 (11%)	84 (76%)	15 (13%)	12 (11%)				0.98
Reading Achievement	229 (92%)	18 (8%)	0 (0%)	102 (94%)	7 (6%)	0 (0%)				0.65

Score 85 = normal, Score 71 to 84 = moderate disability, Score 70 = severe disability