



## Original Article

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**Abstract**

**Objectives:** To evaluate the motor proficiency, identify risk factors for abnormal motor scores, and examine the relationship between motor proficiency and health-related quality of life in school-aged patients with CHD. **Study design:** Patients  $\geq 4$  years old referred to the cardiac neurodevelopmental program between June 2017 and April 2020 were included. Motor skills were evaluated by therapist-administered *Bruininks-Oseretsky Test of Motor Proficiency Second-Edition Short Form* and parent-reported *Adaptive Behavior Assessment System* and *Patient-Reported Outcomes Measurement Inventory System Physical Functioning* questionnaires. Neuropsychological status and health-related quality of life were assessed using a battery of validated questionnaires. Demographic, clinical, and educational variables were collected from electronic medical records. General linear modelling was used for multivariable analysis. **Results:** The median motor proficiency score was the 10<sup>th</sup> percentile, and the cohort ( $n = 272$ ; mean age: 9.1 years) scored well below normative values on all administered neuropsychological questionnaires. In the final multivariable model, worse motor proficiency score was associated with family income, presence of a genetic syndrome, developmental delay recognised in infancy, abnormal neuroimaging, history of heart transplant, and executive dysfunction, and presence of an individualised education plan ( $p < 0.03$  for all predictors). Worse motor proficiency correlated with reduced health-related quality of life. Parent-reported adaptive behaviour ( $p < 0.001$ ) and physical functioning ( $p < 0.001$ ) had a strong association with motor proficiency scores. **Conclusion:** This study highlights the need for continued motor screening for school-aged patients with CHD. Clinical factors, neuropsychological screening results, and health-related quality of life were associated with worse motor proficiency.

**Introduction**

CHD is the most common birth defect, affecting approximately one percent of live births.<sup>1</sup> With advances in the surgical and medical management of CHD, two-thirds of children survive well into adulthood.<sup>2</sup> The focus has shifted to addressing the morbidities associated with living long-term with CHD. Neurodevelopmental morbidity is one of the most important long-term issues faced by CHD survivors and is characterised by a pattern of neurodevelopmental deficits that may change over time, including deficits in language, attention, visual-motor integration, working memory, processing speed, executive functioning, behaviour and emotional functioning, social communication, and fine and gross motor skills.<sup>3,4</sup> Long term, this combination of deficits may affect educational achievements, employability, life-long earnings, insurability, and health-related quality of life.<sup>5</sup> Motor deficits specifically warrant attention as they can impact physical and adaptive functioning and quality of life throughout the lifespan.<sup>4,6,7</sup> While delays in motor skill development are commonly identified in early childhood, less is known about school-aged patients with CHD, who face increasingly robust physical demands and expectations set by not only their caregivers and medical providers but also their peer group. This may impact emotional health and well-being and influence future engagement in physical activity, which may in turn affect long-term health outcomes and quality of life.

The aims of this study were 1) To describe the motor outcomes of high-risk school-aged patients with CHD; 2) to identify risk factors (clinical, demographic, neurodevelopmental, and educational) associated with worse motor outcomes; and 3) to identify the association between motor outcomes and health-related quality of life. We hypothesised that school-aged patients with CHD would have below-average motor scores and that lower motor scores would be associated with clinical, demographic, neurodevelopmental, and educational factors. In

addition, we hypothesised that lower motor scores would be associated with lower health-related quality of life scores.

## Materials and methods

### Study design

We conducted a single-centre cross-sectional study utilising patient data from routine clinical care in the cardiac neurodevelopmental program at Ann & Robert H. Lurie Children's Hospital of Chicago. As part of routine nursing intake, patients older than 4 years of age were screened for clinic referral using criteria from the American Heart Association/American Academy of Pediatrics' scientific statement that categorised pediatric CHD patients at high risk for neurodevelopmental deficits<sup>4</sup> in all affiliated cardiology outpatient clinics. Patients were also referred based on provider concern. All patients evaluated in the clinic underwent a battery of screening neurodevelopmental questionnaires, a motor assessment by a physical therapist, and structured interviews with a cardiologist, developmental behavioural pediatrician, social worker, and education specialist. This study was approved by the Institutional Review Board (IRB # 2020-3560).

### Patient population

Patients were included if they participated in a motor evaluation and completed neurodevelopmental, physical functioning, behavioural and emotional, adaptive functioning, and health-related quality of life assessment in the clinic between June 2017 and April 2020.

### Data collection

Demographic, clinical, educational, and therapy utilisation information were collected from the electronic medical record and are shown in Table 1. Zip code was used to determine median household income from existing US Census Data. In subjects with CHD, their underlying anatomy was categorised into biventricular CHD or single ventricle CHD. Educational environment included the type of classroom setting and whether the patient had educational supports. Therapy utilisation such as physical therapy, occupational therapy, and speech and language therapy was recorded.

The primary outcome variable was the Bruininks-Oseretsky Test of Motor Proficiency Second Edition Short Form, which is administered by a trained provider to assess the general motor proficiency for patients aged 4–21 years old.<sup>8,9</sup> Motor results are reported as both percentiles based on age and gender and by categories defined as well-below average ( $\leq 2^{\text{nd}}$  percentile), below average ( $3^{\text{rd}}$ – $17^{\text{th}}$  percentile), and average or above ( $>17^{\text{th}}$  percentile).<sup>8,9</sup>

Neurodevelopmental, physical functioning, behavioural and emotional, adaptive functioning, and health-related quality of life assessment variables were assessed through a battery of age-specific validated questionnaires (Table 2). These included the *Behavior Rating Inventory of Executive Function Parent Form, Preschool or 2<sup>nd</sup> Edition*,<sup>10</sup> *Conners 3<sup>rd</sup> Edition Parent Rating Scales*,<sup>11</sup> *Patient-Report Outcomes Measurement Information System Physical Functioning Parent Proxy Report* and *Pediatric Self-Report*,<sup>12</sup> *Behavior Assessment System for Children Parent Scales* and *Self-Report Rating Scales, 3<sup>rd</sup> Edition*,<sup>13</sup> *Adaptive Behavior Assessment System Parent/Primary Caregiver Form, 3<sup>rd</sup> Edition*,<sup>14</sup> *Pediatric Quality of Life Inventory Parent-Proxy Report*

and *Child-Self Report*,<sup>15</sup> and *Pediatric Cardiac Quality of Life Index Child Form* and *Parent Form*.<sup>16</sup>

### Statistical analyses

Data distribution was summarized using measures of central tendency (means and medians) and variability (standard deviations and interquartile range) for continuous variables and frequencies (percent) for dichotomous or categorical variables. To examine associations between motor skills and outcomes, as well as covariates, motor scores were also logarithmically transformed to establish a normal distribution. Univariate associations were tested using bi-serial or Pearson's correlations (depending on variable type). Correlations were interpreted as follows: poor agreement  $\leq 0.20$ , fair agreement 0.21 to 0.40, moderate agreement 0.41 to 0.6, good agreement 0.61 to 0.8, excellent agreement  $\geq 0.81$ .<sup>17</sup> To identify risk factors, two distinct models were created using data from the univariate analysis: 1) Demographic/Clinical Model and 2) Neurodevelopmental/Educational Model. Demographic and clinical variables were included in the Demographic/Clinical Model if they were associated with the log transformed motor score at a  $p < 0.15$  level. Dichotomous predictors that had less than 5% in one category were excluded from the multivariable analysis. In the Neurodevelopmental/Educational Model only included parent-reported neurodevelopmental measures and educational variables. Neurodevelopmental, physical functioning, behavioural and emotional, adaptive functioning, and health-related quality of life assessment variables measures included multiple inter-related domains; therefore, the measure having the highest correlation with the log transformed motor score was chosen for the multivariable analysis. The final model combined significant demographic and clinical variables with significant neurodevelopmental and educational variables. General linear modelling was used to test the association between demographic, clinical, educational, and neurodevelopmental measures and log transformed motor scores. Effect size reported as a Partial Eta<sup>2</sup> was presented to show group scores for significant variables. Model fit statistics including R<sup>2</sup> were also presented. All analysis was conducted using SAS 9.4©.

## Results

### Patient population

Of the 272 patients who met inclusion criteria, 50.4% were male ( $n = 127$ ), with a mean age of  $9.1 \pm 3.5$  years (Table 1). The type of heart disease at original diagnosis was CHD in 89% of patients and cardiomyopathy in 9% of patients. Patient's type of heart disease did change over time based on the need for heart transplantation. At the time of clinic evaluation, the type of heart disease was single ventricle CHD in 25% ( $n = 66$ ), biventricular CHD in 56% ( $n = 150$ ), transplanted heart in 19% ( $n = 51$ ), cardiomyopathy without transplant 0.8% ( $n = 2$ ), and other 1.1% ( $n = 3$ ). For those with heart transplant, the heart disease at original diagnosis was split nearly evenly between cardiomyopathy and single ventricle [49% ( $n = 25$ ) vs. 43% ( $n = 22$ )]. Open heart surgery in the first year of life was the most common reason for referral to the neurodevelopmental clinic (77.2%).

The median motor score was 10<sup>th</sup> percentile (interquartile range: 1–19). Overall, 190 patients (69.9%) scored below average [ $n = 123$  (45.2%)] or well-below average [ $n = 67$  (24.6%)] for motor proficiency (Figure 1). Motor scores for patients with an original diagnosis of biventricular and single ventricle CHD were

**Table 1.** Characteristics of the cohort

	<i>n</i> (%)	Correlation with log transformed BOT-2 ( <i>p</i> -value)
<b>Total cohort (<i>n</i> = 272)</b>		
<b>Demographic variables</b>		
Age at visit, mean (SD)	9.1 (3.5)	-0.06 (0.347)
Male	137 (50.4)	-0.03 (0.565)
Race		
American Indian/ Alaskan Native	1 (0.4)	n/a
Asian	16 (5.9)	0.03 (0.641)
Black/African-American	29 (10.7)	-0.05 (0.366)
White	128 (47.1)	0.04 (0.533)
Other	96 (35.3)	-0.02 (0.746)
Declined	2 (0.7)	n/a
Hispanic/Latino(a) Ethnicity	117 (43.0)	-0.04 (0.472)
Mean Household Income by Census Tract (SD)	\$66,377 (27,545)	0.17 (0.004)
Married	196 (74.8)	0.09 (0.140)
Private Insurance	120 (46.2)	0.16 (0.009)
<b>Clinical Variables</b>		
<i>Type of Heart Disease at Original Diagnosis<sup>b</sup></i>		
CHD	242 (89.0)	
Single Ventricle	88 (32.4)	
Biventricular	154 (67.6)	
Cardiomyopathy	27 (9.3)	
Other <sup>a</sup>	3 (1.1)	
<i>Type of Heart Disease at Clinic Evaluation<sup>b</sup></i>		
Single Ventricle CHD	66 (25)	
Biventricular CHD	150 (56)	
Transplanted Heart	51 (19)	
Cardiomyopathy without Transplant	2 (0.8)	
Other <sup>a</sup>	3 (1.1)	
<i>Reason for Referral</i>		
Open Heart Surgery in the First Year of Life	210 (77.2)	
Unrepaired Cyanotic Heart Lesion	10 (3.7)	
Provider Concern	52 (0.19)	
<i>Risk Factors</i>		
History of Prematurity	32 (11.8)	-0.16 (0.008)
DD Recognized in Infancy	138 (50.7)	-0.43 (<0.001)
Genetic Syndrome Associated with DD	51 (19.7)	-0.36 (<0.001)
History of ECMO or VAD	30 (11.0)	-0.07 (0.268)
History of Heart Transplant	51 (8.7)	-0.10 (0.108)
Previous Exposure to CPR	33 (12.1)	-0.13 (0.037)
Perioperative Length of Stay >14 Days	191 (70.2)	-0.03 (0.602)
History of Perioperative Seizures	13 (4.8)	-0.14 (0.024)
History of Abnormal Head Imaging or Microcephaly	36 (12.2)	-0.26 (<0.001)
Activity Restricted by Cardiologist	40 (14.8)	0.04 (0.510)

(Continued)

Table 1. (Continued)

	<i>n</i> (%)	Correlation with log transformed BOT-2 ( <i>p</i> -value)
<b>Educational Variables</b>		
Educational Environment <sup>d</sup>		0.41 (<0.001)
Traditional Classroom Setting	89 (32.7)	
Classroom Setting with Specialized Services	164 (60.3)	
Alternative Placement	19 (7.0)	
Educational Supports <sup>e</sup>		-0.39 (<0.001)
Individualized Education Program	135 (82.8)	
504 Plan	23 (14.1)	
Other	5 (3.1)	
<b>Therapy Utilization</b>		
Current Use of Augmentative Communication	18 (6.6)	-0.32 (<0.001)
History of Receiving Speech and Language Therapy	205 (75.4)	-0.27 (<0.001)
Current Use of Adaptive Technology	17 (6.2)	-0.22 (<0.001)
Currently Receiving PT Services	49 (18.0)	-0.46 (<0.001)
History of Previously Receiving PT Services	174 (64.0)	-0.19 (0.002)
Currently Receiving OT Services	83 (30.5)	-0.52 (<0.001)
History of Previously Receiving OT Services	180 (66.2)	-0.26 (<0.001)

CPR = cardiopulmonary resuscitation; DD = developmental delay; ECMO = extracorporeal membrane oxygenation; IEP [Individualized Education Plan]; OT = occupational therapy; PT = physical therapy; VAD [ventricular assist device]; SD = standard deviation. <sup>a</sup>Type of heart disease diagnosis was endocarditis (*n* = 1) and arrhythmia (*n* = 2). <sup>b</sup>Neither Type of Heart Disease at Original Diagnosis nor Type of Heart Disease at Clinic Evaluation were associated with the log transformed BOT-2 score. <sup>d</sup>Traditional Classroom Setting used as reference value. <sup>e</sup>Individualized Education Program used as reference value.

both similarly low and not significantly different. However, patients with single ventricle CHD who had undergone heart transplantation had the worst motor performance (4<sup>th</sup> percentile) in comparison to patients with an original diagnosis of biventricular CHD who had not undergone heart transplantation (10<sup>th</sup> percentile) ( $p = 0.029$ ). Outcomes of the neurodevelopmental questionnaires, normative values, and the association with log transformed motor scores are described in Table 2. Questionnaires indicated lower than average functioning compared to published normative values in measures of adaptive functioning, executive functioning, attention and hyperactivity, emotional health, and quality of life.

### Risk factors for motor deficits

The demographic, clinical, educational, and therapy utilisation variables associated with the log transformed motor scores on univariate analysis are found in Table 1. Univariate associations between neurodevelopmental variables and log transformed motor scores are found in Table 2. In the Demographic/Clinical Model, family income, suspected genetic syndrome, developmental delay recognised in infancy, abnormal neuroimaging, and history of heart transplantation were associated with log transformed motor scores; explaining 30% of the variation in log transformed motor scores. In the Neurodevelopmental/Educational model, the Generalized Executive Composite domain of the Behavior Rating Inventory of Executive Function Parent Form, 2<sup>nd</sup> Edition and presence of an Individualized Education Program were associated with log transformed motor scores, explaining 21% of the variation in log transformed motor scores. In the final

combined multivariable model, family income, suspected genetic syndrome, developmental delay recognised in infancy, abnormal neuroimaging, history of heart transplantation, Behavior Rating Inventory of Executive Function Parent Form, 2<sup>nd</sup> Edition Global Executive Composite, and presence of an Individualized Education Program explained 35% of the variation in log transformed motor outcomes (Table 3).

### Motor outcomes and quality of life

Health-related quality of life was measured for children  $\geq 8$  years of age using the Pediatric Cardiac Quality of Life Inventory. Parent-proxy and patient-reported health-related quality of life Total, Disease Impact, and Psychosocial Impact sub-scale scores were lower than scores published for patients with mild CHD. Motor performance had a fair association with Pediatric Cardiac Quality of Life Inventory parent-proxy Disease Impact score ( $r = 0.30$ ,  $p = 0.002$ ) and with patient-reported Disease Impact score ( $r = 0.20$ ,  $p = 0.047$ ). Neither parent-proxy nor patient-reported Pediatric Cardiac Quality of Life Inventory Total or Psychosocial Impact scores were correlated with motor performance.

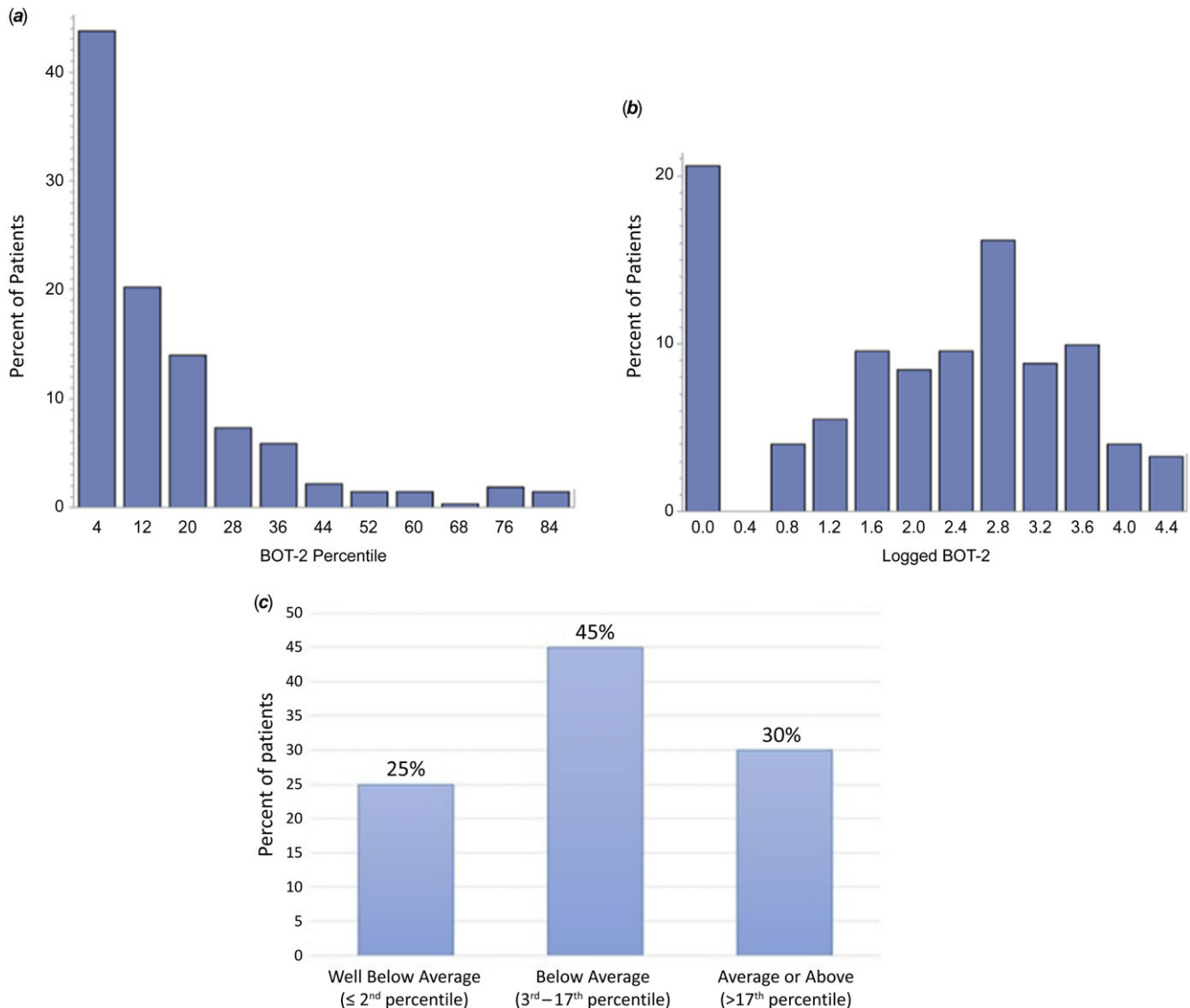
### Discussion

The purpose of this study was to describe the motor proficiency of school-aged children born with CHD, to identify risk factors that may result in worse motor outcomes, and to assess the association of motor scores with health-related quality of life. This study is one of only a few to assess motor outcomes beyond early childhood and confirms that motor deficits persist well into school-aged years. In

**Table 2.** Summary statistics for neurodevelopmental questionnaires

Specific subscale		Normative scores		Clinic populations scores		t Value	Pr >  t	Correlation with log transformed BOT r (p value)
		Mean	SD	Mean	SD			
Parent-Proxy Reported Questionnaires								
Behavior Rating Inventory of Executive Function Parent Form, 2 <sup>nd</sup> Edition (BRIEF)	Behavior Regulation Index (n = 242)	50	10	55.0	12.5	-5.17	<0.001	-0.26 (<0.001)
	Metacognition Index (n = 242)	50	10	58.1	12.3	-8.27	<0.001	-0.28 (<0.001)
	Global Executive Composite (n = 242)	50	10	58.1	12.3	-8.27	<0.001	-0.30 (<0.001)
Conners 3 <sup>rd</sup> Edition Parent Rating Scales	Inattention (n = 196)	50	10	64.8	15.7	-11.73	<0.001	-0.23 (0.001)
	Hyperactivity (n = 196)	50	10	61.6	16.0	-9.06	<0.001	-0.12 (0.100)
	ADHD Index (n = 196)	50	10	53.8	33.8	-1.53	0.127	-0.21 (0.004)
	Global Index (n = 196)	50	10	61.2	15.1	-9.15	<0.001	-0.19 (0.01)
Patient-Reported Outcomes Measure Information System, Proxy Report (PROMIS)	Fatigue Index (n = 225)	50	10	46.5	9.7	4.02	<0.001	-0.19 (0.001)
	Cognition Function Index (n = 230)	50	10	46.0	9.3	4.67	<0.001	0.22 (0.001)
	Physical Function Index (n = 230)*	50	10	46.7	8.7	3.78	<0.001	0.54 (<0.001)
Behavior Assessment System for Children Parent Scales, 3 <sup>rd</sup> Edition (BASC)	Externalizing Problems (n = 247)	50	10	51.4	10.1	-1.62	0.105	-0.01 (0.913)
	Internalizing Problems (n = 247)	50	10	52.3	11.7	-2.44	0.015	0.06 (0.322)
	Behavior Symptom Index (n = 246)	50	10	54.0	11.0	-4.44	<0.001	-0.07 (0.309)
	Adaptive Skills (n = 247)	50	10	43.5	10.7	7.33	<0.001	0.20 (0.002)
Adaptive Behavior Assessment System, Parent/Primary Caregiver Form 3 <sup>rd</sup> Edition (ABAS)	General Adaptive Composite (n = 212)*	100	15	86.1	17.4	9.66	<0.001	0.59 (<0.001)
	Practical (n = 213)*	100	15	87.0	17.4	8.98	<0.001	0.57 (<0.001)
Pediatric Quality of Life Inventory Parent-Proxy Report (Peds QL)	Cognitive Fatigue (n = 229)	90.1 <sup>a30</sup>	14.7	55.8	26.9	-15.19	<0.001	0.27 (<0.001)
Pediatric Cardiac Quality of Life Index Parent-Proxy Form (PCQLI <sup>b</sup> )	Total (n = 118)	82.4 <sup>c31</sup>	16.8	64.8	16.5	-6.46	<0.001	0.19 (0.052)
	Disease Impact (n = 118)*	41.2 <sup>c31</sup>	9.2	30.5	8.7	-4.6	<0.001	0.30 (0.002)
	Psychosocial Impact (n = 119)	41.1 <sup>c31</sup>	8.2	34.5	9.4	-4.44	<0.001	0.06 (0.526)
Patient-Reported Questionnaires								
Patient-Reported Outcomes Measure Information System (PROMIS)	Fatigue Index (n = 112)	50	10	47.6	10.2	2.16	0.032	-0.13 (0.171)
	Cognition Function Index (n = 114)	50	10	45.3	8.0	4.97	<0.001	0.19 (0.041)
Behavior Assessment System for Children (BASC)	Internalizing Problems (n = 118)	50	10	50.5	10.8	-0.45	0.653	-0.11 (0.207)
	Hyperactivity (n = 116)	50	10	49.2	9.4	0.74	0.457	-0.01 (0.912)
	Personal Adjustment (n = 117)	50	10	52.3	11.1	-2.04	0.042	0.02 (0.783)
Pediatric Quality of Life Inventory (PedsQL)	Cognitive Fatigue (n = 116)	82.1 <sup>a30</sup>	17	58.5	26.0	-8.19	<0.001	0.17 (0.066)
Pediatric Cardiac Quality of Life Index (PCQLI <sup>b</sup> )	Total (n = 111)	83 <sup>c31</sup>	14.2	66.2	16.8	-6.33	<0.001	0.11 (0.244)
	Disease Impact (n = 112)	41.5 <sup>c31</sup>	7.4	31.7	8.2	-7.44	<0.001	0.20 (0.047)
	Psychosocial Impact (n = 114)	41.5 <sup>c31</sup>	8.1	34.7	9.5	-4.54	<0.001	0.06 (0.56)

\*Indicates statistically significant correlation with log transformed BOT-2 Score. Self-Report questionnaires have significantly fewer responses, so were not included in the final analysis. aVarni JW, Limbers CA, Sorensen LG, et al. PedsQL™ Cognitive Functioning Scale in pediatric liver transplant recipients: feasibility, reliability, and validity. Qual Life Res. 2011;20(6):913-921. doi:10.1007/s11136-010-9823-1. bPCQLI administered to patients and parent of patients 8 to 18 years of age. cNormative scores based on patients with Mild CHD.



**Figure 1.** (a) Distribution of Bruininks-Oseretsky Test of Motor Proficiency Second Edition Short Form (BOT-2) scores by percentile. (b) Log transformed distribution of BOT-2 scores. (c) BOT-2 Performance organized by category.

our cohort, 70% of patients scored either below or well-below average on motor proficiency assessment. Worse motor proficiency was significantly associated with family income, suspected genetic syndrome, developmental delay recognised in infancy, abnormal imaging, history of a heart transplant, executive functioning, and having an Individualised Education Program. Worse outcomes on motor assessment were also associated with lower health-related quality of life in the disease impact domain. These outcomes highlight the need for dedicated long-term motor and adaptive function support, including physical therapy and occupational therapy in the CHD population.

It has been well-established that infants and toddlers with CHD have deficits in fine and gross motor skills, and that the degree of impairment may change over time.<sup>18,19</sup> Mussatto et al described that 75% of patients ages 5.5–37 months scored in the “at risk” or “delayed” range in 1 or more domains on standardised neuro-developmental testing and even more were found to be delayed with ongoing surveillance.<sup>18</sup> To further emphasise the value of longitudinal evaluation, Brosig et al compared scores at two and

four years of age and found that the number of children who were classified as delayed or at-risk increased between two and four years of age for both cognitive and fine motor skills.<sup>19</sup> This critical period of development corresponds to the time when children transition out of early intervention programs, highlighting the need for ongoing follow-up to ensure a smooth transition to early childhood programs or outpatient therapies.

Despite numerous early childhood studies demonstrating an evolution of motor delays over time,<sup>20,21</sup> data in the school-aged and adolescent population are more limited. In a small cohort of 33 patients who underwent cardiac surgery in the first year of life, 41% of 5-year-old patients demonstrated below-average motor proficiency on the Bruininks-Oseretsky Test of Motor Proficiency Second Edition Short Form.<sup>22</sup> A larger study of 233 congenital heart surgery survivors investigated the motor assessment of 6-year-old patients utilising the Zurich Neuromotor Assessment.<sup>23</sup> This study similarly found that children with CHD scored lower on motor testing, with dynamic balance most significantly affected. In a systematic review by Bolduc et al, only 11% of studies examined

**Table 3.** Multivariable models for logged BOT scores

Demographic and Clinical Variables	Partial Eta-Square	Chi-Square	Pr > ChiSq
Family Income	0.01	4.09	0.043
Suspected Genetic Syndrome	0.08	15.81	<0.001
Development Delay Recognized in Infancy	0.11	29.27	<0.001
Abnormal Neuroimaging	0.03	8.62	0.003
History of a Heart Transplantation	0.02	5.72	0.017
Model fit statistics (df: 5,266; f: 28.68, $p < 0.001$ ; $R^2$ : 0.30)			
Neurodevelopmental and Educational Variables	Partial Eta-Square	Chi-Square	Pr > ChiSq
Behavior Rating Inventory of Executive Function Parent Form, 2 <sup>nd</sup> Edition Global Executive Composite	0.06	14.59	<0.001
Presence of an Individualized Education Program	0.13	33.49	<0.001
Model fit statistics (df: 2,239; f: 31.32, $p < 0.001$ ; $R^2$ : 0.21)			
Final Model	Partial Eta-Square	Chi-Square	Pr > ChiSq
Family Income	0.02	5.44	0.020
Suspected Genetic Syndrome	0.03	7.49	0.006
Development Delay Recognized in Infancy	0.06	15.37	<0.001
Abnormal Neuroimaging	0.02	4.77	0.029
History of a Heart Transplantation	0.02	10.49	0.001
Behavior Rating Inventory of Executive Function Parent Form, 2 <sup>nd</sup> Edition Global Executive Composite	0.04	4.23	0.040
Presence of an Individualized Education Program	0.05	12.13	<0.001
Model fit statistics (df: 7,234; f: 17.84, $p < 0.001$ ; $R^2$ : 0.35)			

motor skills in the school-aged or older CHD population, with the majority of studies being completed in Europe in an earlier surgical era.<sup>7</sup> Our cohort is the largest study of school-aged CHD patients in the United States and the average age of 9.1 years (standard deviation = 3.5) offers more longitudinal insight into the ongoing motor deficiencies of a heterogeneous contemporary cohort of children with CHD. The results identify a larger proportion of children with significant motor deficits compared to other studies. Alarmingly, despite the overwhelming presence of motor delays, only 18% and 31% of children in our cohort were receiving physical therapy and occupational therapy, respectively, at the time of evaluation, suggesting that rehabilitation and developmental services may be under-utilised in this population. Notably, the Individuals with Disabilities Education Act mandate focuses on services that improve access to the educational environment rather than on maximising functional outcomes, which may limit the ability of children with CHD to qualify for school-based services despite ongoing motor concerns. A recent study by Wehrle et al reinforced the idea that neurodevelopmental therapy awareness may be reduced in children with CHD. Despite facing similar developmental challenges and performance compared to very preterm infants, patients with CHD were significantly less likely to receive motor-related therapy services.<sup>24</sup>

We also demonstrated that a combination of socio-economic, psychological, and educational factors predicted worse motor performance in our cohort, which underscores the importance of multifactorial screening and highlights an opportunity for multiple targeted interventions in children with CHD. On multivariable analysis, lower income, presence of an Individualised Education Program, and executive dysfunction predicted worse motor

performance. Several previous studies have also identified socio-economic status as predictive of motor scores in univariate analysis.<sup>23,25</sup> Similarly, an association between educational environment and motor outcomes has been identified by Liamlahi et al, which identified children with higher rates of behavioural abnormalities and motor problems requiring higher rates of special education.<sup>26</sup> In our study, worse executive function, as measured by the Behavior Rating Inventory of Executive Function Parent Form, 2<sup>nd</sup> Edition Global Executive Composite, was associated with worse motor outcomes, perhaps related to difficulties with motor planning required to complete the testing. These results highlight the value of neuropsychological assessment in patients with CHD, as performance on specific sub-tests and overall school performance may also help identify individuals who would benefit from additional motoric evaluation and potential intervention.

The finding of decreased health-related quality of life scores in the high-risk school-aged CHD population and its association with lower motor scores further emphasises the importance of regular motor screening and intervention in this population. Compared to the published reference population with mild CHD, the cohort of patients followed in our neurodevelopmental clinic had worse health-related quality of life as measured by the Pediatric Cardiac Quality of Life Inventory parent-proxy and self-report forms. Furthermore, the disease-impact score on both the parent-proxy and self-report Pediatric Cardiac Quality of Life Inventory was associated with lower motor scores. The complex interplay between motor performance, executive function, and health-related quality of life has been explored in previous studies. Marino et al identified that gross motor ability and executive function and

mood were predictors of lower health-related quality of life scores.<sup>27</sup> Similarly, Mellion et al found that health-related quality of life, specifically physical functioning, was lower in patients with CHD compared with healthy controls.<sup>28</sup> Based on the results of these studies, additional research may be warranted to target motor outcomes as a potential intervention strategy aimed at improving health-related quality of life.

Overall, the findings of our paper support the published Cardiac Neurodevelopmental Outcomes Collaborative guidelines, which promote multi-dimensional screening for patients with CHD, ideally in a dedicated multidisciplinary clinic.<sup>29</sup> Comprehensive screening and intervention may be important tools to improve the neurodevelopmental outcomes and improve health-related quality of life in this population. Based on the high rate of motor dysfunction, motor screening needs to be included.

A strength of our study is the large sample of at-risk school-aged patients, a group whose motor performance has historically received limited attention.<sup>7</sup> However, this study has several limitations. Patients with more severe developmental difficulties may be both more likely to be referred to the cardiac neurodevelopmental clinic and more likely to complete the evaluation. Additionally, because gross and fine motor skills were categorised together, the overall motor deficiency of our cohort may be biased due to the lack of differentiation between the domains. Future studies are needed to track the longitudinal progress of patients, to categorise motor outcomes based on specific skills, and to develop intervention strategies that target motor outcomes.

## Conclusion

In conclusion, motor deficits in patients with CHD persist beyond early childhood and are associated with worse health-related quality of life. Predictors of adverse motor scores include family income, suspected genetic syndrome, developmental delay recognised in infancy, abnormal imaging, history of a heart transplant, executive dysfunction, and presence of an Individualized Education Program. The clinical, psychosocial, and educational factors offer potential targets for intervention and highlight the importance of multi-disciplinary support and multi-dimensional screening for patients with CHD who are deemed high risk.

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## References

1. Wu W, He J, Shao X. Incidence and mortality trend of congenital heart disease at the global, regional, and national level, 1990–2017. *Medicine (Baltimore)* 2020; 99: e20593. DOI: [10.1097/MD.00000000000020593](https://doi.org/10.1097/MD.00000000000020593).
2. Marelli AJ, Ionescu-Ittu R, Mackie AS, Guo L, Dendukuri N, Kaouache M. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. *Circulation* 2014; 130: 749–756. DOI: [10.1161/CIRCULATIONAHA.113.008396](https://doi.org/10.1161/CIRCULATIONAHA.113.008396).
3. Verrall CE, Blue GM, Loughran-Fowlds A, et al. 'Big issues' in neurodevelopment for children and adults with congenital heart disease. *Open Heart* 2019; 6: e000998. DOI: [10.1136/openhrt-2018-000998](https://doi.org/10.1136/openhrt-2018-000998).
4. Marino BS, Lipkin PH, Newburger JW, et al. Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association. *Circulation* 2012; 126: 1143–1172. DOI: [10.1161/CIR.0b013e318265ee8a](https://doi.org/10.1161/CIR.0b013e318265ee8a).
5. Ilardi D, Ono KE, McCartney R, Book W, Stringer AY. Neurocognitive functioning in adults with congenital heart disease. *Congenit Heart Dis* 2017; 12: 166–173. DOI: [10.1111/chd.12434](https://doi.org/10.1111/chd.12434).
6. Reuben DB, Magasi S, McCreath HE, et al. Motor assessment using the NIH toolbox. *Neurology* 2013; 80: S65–75. DOI: [10.1212/WNL.0b013e3182872e01](https://doi.org/10.1212/WNL.0b013e3182872e01).
7. Bolduc ME, Dionne E, Gagnon I, Rennick JE, Majnemer A, Brossard-Racine M. Motor impairment in children with congenital heart defects: a systematic review. *Pediatrics* 2020; 146: e153–e639. DOI: [10.1542/peds.2020-0083](https://doi.org/10.1542/peds.2020-0083).
8. Bruininks RH, Bruininks BD. BOT2: Bruininks-Oseretsky test of motor proficiency. Pearson, Assessments, 2005.
9. Lucas BR, Latimer J, Doney R, et al. The Bruininks-Oseretsky test of motor proficiency-short form is reliable in children living in remote Australian aboriginal communities. *BMC Pediatr* 2013; 13: 135. DOI: [10.1186/1471-2431-13-135](https://doi.org/10.1186/1471-2431-13-135).
10. Gioia GA, Isquith PK, Guy SC, Kenworthy L. BRIEF-2: Behavior rating inventory of executive function. Psychological Assessment Resources, Lutz, FL, 2015.
11. Conners CK. Conners, 3rd edition. Multi-Health Systems Toronto, 2008.
12. Cella D, Riley W, Stone A, et al. The Patient-Reported Outcomes Measurement Information System (PROMIS) developed and tested its first wave of adult self-reported health outcome item banks: 2005–2008. *J Clin Epidemiol* 2010; 63: 1179–1194. DOI: [10.1016/j.jclinepi.2010.04.011](https://doi.org/10.1016/j.jclinepi.2010.04.011).
13. Reynolds C, Kamphaus R. Behavior assessment system for children-Third Edition (BASC-3). Pearson, Bloomington, MN, 2015.
14. Harrison PL, Oakland T. ABAS-3: Adaptive behavior assessment system. Western Psychological Services Los, Angeles, CA, 2015.
15. Varni JW, Seid M, Rode CA. The PedsQL: measurement model for the pediatric quality of life inventory. *Med Care* 1999; 37: 126–139. DOI: [10.1097/00005650-199902000-00003](https://doi.org/10.1097/00005650-199902000-00003).
16. Marino BS, Shera D, Wernovsky G, et al. The development of the pediatric cardiac quality of life inventory: a quality of life measure for children and adolescents with heart disease. *Qual Life Res* 2008; 17: 613–626. DOI: [10.1007/s11136-008-9323-8](https://doi.org/10.1007/s11136-008-9323-8).
17. Landis JR, Koch GG. The measurement of observer agreement for categorical data. *Biometrics* 1977; 33: 159–174.
18. Mussatto KA, Hoffmann RG, Hoffman GM, et al. Risk and prevalence of developmental delay in young children with congenital heart disease. *Pediatrics* 2014; 133: e570–e577. DOI: [10.1542/peds.2013-2309](https://doi.org/10.1542/peds.2013-2309).
19. Brosig CL, Bear L, Allen S, et al. Neurodevelopmental outcomes at 2 and 4 years in children with congenital heart disease. *Congenit Heart Dis* 2018; 13: 700–705. DOI: [10.1111/chd.12632](https://doi.org/10.1111/chd.12632).
20. Sprong MC, van Brussel M, de Vries LS, et al. Longitudinal motor-developmental outcomes in infants with a critical congenital heart defect. *Children* 2022; 9: 570.
21. Sprong MC, Huijgen BC, de Vries LS, et al. Early determinants of adverse motor outcomes in preschool children with a critical congenital heart defect. *J Clin Med* 2022; 11: 5464.
22. Long SH, Eldridge BJ, Harris SR, Cheung MM. Motor skills of 5-year-old children who underwent early cardiac surgery. *Cardiol Young* 2016; 26: 650–657. DOI: [10.1017/S1047951115000797](https://doi.org/10.1017/S1047951115000797).
23. Naef N, Liamlahi R, Beck I, et al. Neurodevelopmental profiles of children with congenital heart disease at school age. *J Pediatr* 2017; 188: 75–81. DOI: [10.1016/j.jpeds.2017.05.073](https://doi.org/10.1016/j.jpeds.2017.05.073).
24. Wehrle FM, Barta T, Adams M, et al. Similarities and differences in the neurodevelopmental outcome of children with congenital heart disease and



- children born very preterm at school entry. *J Pediatr* 2022; 250: 29–37 e1. DOI: [10.1016/j.jpeds.2022.05.047](https://doi.org/10.1016/j.jpeds.2022.05.047).
25. Bucholz EM, Sleeper LA, Goldberg CS, et al. Socioeconomic status and long-term outcomes in single ventricle heart disease. *Pediatrics* 2020;146: e20201240. DOI: [10.1542/peds.2020-1240](https://doi.org/10.1542/peds.2020-1240).
  26. Liamlahi R, von Rhein M, Buhner S, et al. Motor dysfunction and behavioural problems frequently coexist with congenital heart disease in school-age children. *Acta Paediatr* 2014; 103: 752–758. DOI: [10.1111/apa.12639](https://doi.org/10.1111/apa.12639).
  27. Marino BS, Cassidy A, Drotar D, Wray J. The impact of neurodevelopmental and psychosocial outcomes on health-related quality of life in survivors of congenital heart disease. *J Pediatr* 2016; 174: 11–22 e2. DOI: [10.1016/j.jpeds.2016.03.071](https://doi.org/10.1016/j.jpeds.2016.03.071).
  28. Mellion K, Uzark K, Cassidy A, et al. Health-related quality of life outcomes in children and adolescents with congenital heart disease. *J Pediatr* 2014; 164: 781–788 e1. DOI: [10.1016/j.jpeds.2013.11.066](https://doi.org/10.1016/j.jpeds.2013.11.066).
  29. Ilardi D, Sanz JH, Cassidy AR, et al. Neurodevelopmental evaluation for school-age children with congenital heart disease: recommendations from the cardiac neurodevelopmental outcome collaborative. *Cardiol Young* 2020; 30: 1623–1636. DOI: [10.1017/S1047951120003546](https://doi.org/10.1017/S1047951120003546).
  30. Varni JW, Limbers CA, Sorensen LG, et al. PedsQL cognitive functioning scale in pediatric liver transplant recipients: feasibility, reliability, and validity. *Qual Life Res* 2011; 20: 913–921. DOI: [10.1007/s11136-010-9823-1](https://doi.org/10.1007/s11136-010-9823-1).
  31. Czosek RJ, Kaltman JR, Cassidy AE, et al. Quality of life of pediatric patients with long QT syndrome. *Am J Cardiol* 2016; 15: 605–610. DOI: [10.1016/j.amjcard.2015.11.051](https://doi.org/10.1016/j.amjcard.2015.11.051).
  33. Varni JW, Seid M, Rode C. PedsQL. *Measurement model for the pediatric quality of life inventory Version*. 1184–1196. 1998;4.