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Executive Functioning and School Performance among Pediatric Survivors of Complex Congenital Heart Disease

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Objective To investigate the presence and severity of real-world impairments in executive functioning—responsible for children's regulatory skills (metacognition, behavioral regulation)—and its potential impact on school performance among pediatric survivors of complex congenital heart disease (CHD).

Study design Survivors of complex CHD aged 8-16 years (n = 143) and their parents/guardians from a regional CHD survivor registry participated (81% participation rate). Parents completed proxy measures of executive functioning, school competency, and school-related quality of life (QOL). Patients also completed a measure of school QOL and underwent IQ testing. Patients were categorized into 2 groups based on heart lesion complexity: 2-ventricle or single-ventricle.

Results Survivors of complex CHD performed significantly worse than norms for executive functioning, IQ, school competency, and school QOL. Metacognition was more severely affected than behavioral regulation, and metacognitive deficits were more often present in older children. Even after taking into account demographic factors, disease severity, and IQ, metacognition uniquely and strongly predicted poorer school performance. In exploratory analyses, patients with single-ventricle lesions were rated as having lower school competency and school QOL, and patients with 2-ventricle lesions were rated as having poorer behavioral regulation.

Conclusions Survivors of complex CHD experience greater executive functioning difficulties than healthy peers, with metacognition particularly impacted and particularly relevant for day-to-day school performance. Especially in older children, clinicians should watch for metacognitive deficits, such as problems with organization, planning, self-monitoring, and follow-through on tasks. *(J Pediatr 2016;173:154-9)*.

See editorial, p 7

Congenital heart disease (CHD) is the most common congenital structural defect, occurring in 9 out of 1000 live births,[1] with one-third having complex CHD requiring intervention during the neonatal period.[2] At one time, rapid deterioration and death were the expected outcomes for complex CHD, but medical advances have improved 20-year survival rates to around 80%.[3] Unfortunately, survivors often suffer neurodevelopmental morbidity because of prenatal factors (eg, reduced brain volume) as well as chronic or intermittent hypoxia, hypoperfusion, reperfusion, or ischemia related to their circulatory abnormalities and associated medical interventions.[4-7] Accordingly, children with CHD are at higher risk of cognitive deficits and poorer school outcomes than their healthy counterparts,[8-11] with many requiring special educational services.[12,13] In one large multicenter study, 35% of children with CHD required some form of special education.[14]

To better address scholastic concerns, it is important to understand the underlying contributors. Intelligence is a key factor, but IQ accounts for only 25% of variance in school performance for survivors of CHD.[9] Beyond IQ, executive functioning has emerged as an important predictor of school performance in healthy children.[15-17] Executive functioning broadly encompasses 2 main components: behavior regulation (control of emotional and behavioral impulses) and metacognition (planning, organization, self-monitoring, initiation, and follow-through).[18] These executive skills are integral to a child's cognitive, emotional, and social development.

Among pediatric survivors of complex CHD, few studies have examined deficits on structured tests of executive functioning or its real-world implications.

BRIEF	Behavior Rating Inventory of Executive Function
CBCL	Child Behavior Checklist
CCHMC	Cincinnati Children's Hospital Medical Center
CHD	Congenital heart disease
Peds-QL	Pediatric Quality of Life Inventory
QOL	Quality of life

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Most studies indicate roughly one-quarter to one-half of survivors score abnormally low,[8,19,20] although one study found rates as high as three-quarters of survivors.[21] However, these studies generally failed to include patients' self-report or parent proxy measures of executive functioning. Because structured tests can fail to appreciate executive functioning deficits that are evident in real-world settings (eg, trouble starting homework, overreacting to minor infractions),[18] actual rates of impairment may be even greater. In light of growing numbers of survivors of CHD and the potential for long-term morbidity, it is important to understand the risks for real-world executive functioning deficits as well as their impact on what is arguably the most important "job" of school-age children: school performance.

The current study had 2 main aims. The first aim was to investigate whether executive functioning deficits in daily life (per parent report) are differentially present in pediatric survivors of complex CHD surgery and, concurrently, to confirm the presence of lower intellectual functioning and greater school problems. We hypothesized that survivors would have significantly worse group scores and have higher rates of impairment, than published norms across measures of executive functioning, intelligence, and school functioning. Exploratory analyses also examined whether the behavior regulation vs metacognitive aspects of executive functioning were differentially affected and whether effects varied by age or heart lesion complexity (1- vs 2-ventricle disease). Patients with single-ventricle lesions often have many more medical complications because of the complexity of their disease and required treatments. They require multiple operations during their lifetime and have a much higher use of medical care utilization including, the need for cardiac catheterization, multiple hospitalizations, the requirement of multiple medications, and many doctor visits annually. The second aim was to clarify the scholastic implications of executive functioning morbidity in survivors of complex CHD surgery. We hypothesized that poor executive functioning would be associated with worse school functioning, over and above associations with demographic characteristics, intelligence, and heart lesion complexity.

Methods

This cross-sectional study was approved by the Institutional Review Board at Cincinnati Children's Hospital Medical Center (CCHMC). English-speaking survivors of CHD surgery aged 8-16 years were recruited via mailings and phone calls from 2008 and 2010 to a regional registry of children who had all undergone cardiac surgery in the neonatal period (28 days or less) or infancy (1 month to 1 year). Survivors were eligible to participate if they had a primary diagnosis of complex 2-ventricle CHD transposition of the great arteries or tetralogy of Fallot or single-ventricle CHD requiring Fontan palliation, and had undergone cardiac surgery at CCHMC or were followed at CCHMC or Dayton Children's Hospital. Survivors were excluded for other significant medical diseases (eg, cancer); comorbid genetic syndrome contributing to developmental delay; pregnancy; or an unrelated major life event within the past 6 months (eg, serious illness of a family member, parental divorce) that may impact quality of life (QOL). Parents provided written consent, and children provided verbal or written assent. As part of a larger study on neurodevelopmental outcomes, children underwent neuropsychological evaluation while parents completed questionnaires. Demographic information and medical history were obtained via parent report and review of the medical record.

The parent-report version of the Behavior Rating Inventory of Executive Function (BRIEF)[18] is a 86-item questionnaire designed to assess the various facets of executive functioning in an everyday environment in school-aged children aged 5-18 years. The BRIEF has 2 summary indices: metacognition and behavioral regulation. Metacognition encompasses abilities involved in working memory; initiating, planning, and organizing tasks; and monitoring performance. Behavioral regulation is defined as the ability to shift mindset and regulate and control emotions and behavior. Both indices have excellent internal consistency and test-retest reliability[18] and have been shown to be sensitive to executive functioning deficits in patients with surgically corrected CHD.[11] Age- and sex-normed scores are expressed in T-score units, with higher scores indicating worse difficulties.

School performance can be difficult to measure. Grading schemes vary, and academic knowledge (assessed via formal tests) is only indirectly related to day-to-day functioning.[22] With the goal of assessing day-to-day performance at school, we measured the overlapping constructs of school competency and school-related QOL. School competency was assessed by the Child Behavior Checklist (CBCL), which queries parents on the child's typical grades (using a standardized metric) and the need for educational supports.[23] The CBCL is highly reliable and valid, and the School Competency Index is sensitive to adverse effects of CHD.[11] School-related QOL was assessed by the school functioning subscale of the Pediatric Quality of Life Inventory (Peds-QL) version 4.0,[24] a generic health-related QOL measure with scores that are highly reliable, valid, and generalizable[25] and has been used previously in pediatric cardiovascular populations.[10] The Peds-QL school subscale inquires about day-to-day classroom-relevant functional skills (eg, keeping up with schoolwork). Both measures were age- and sex-normed to T-scores, with higher scores indicating better functioning.

The Wechsler Intelligence Scale for Children-Fourth Edition is a widely used, reliable, and valid instrument that assesses intelligence in children aged 6-16 years. To minimize burden, an 8-subtest short form of the Wechsler Intelligence Scale for Children-Fourth Edition was used to yield a prorated full scale IQ.[26]

Demographic covariates included age, sex, race (recoded as white/nonwhite), and yearly family income (dichotomized at $51 000, the US median income level for 2012). The

number of school days missed for any reason (ie, cardiac reasons or for reasons other than cardiac-related) was recoded into number of weeks of school missed (Table I).

Group differences on measures of IQ, executive functioning, and school performance between the entire sample and normative standards as well as by heart lesion complexity were assessed using *t* tests. For more direct clinical relevance, we repeated analyses using χ^2 tests, defining "abnormal" scores on the BRIEF, CBCL, and Peds-QL as those within the worst 10% compared with norms. The relationship between age and outcome variables was assessed using Pearson *r*.

Multivariable analysis was conducted using ordinary least squared regression to determine whether executive functioning impacts school performance above and beyond demographic characteristics, IQ, and heart lesion complexity. A variance inflation score was calculated to assess the level of multicollinearity between the behavioral regulation and metacognition indices in the models. All variables remained in the model in order to test the strength of the relationships between the predictor variables and outcome in the presence of covariates. Parameter and standardized estimates show the change in the relationships between outcome and predictors in raw form (b) as well as the effect size (β) for each variable in the model. An α value of <0.05 was used to determine significance. All analysis was conducted using SAS 9.2 (SAS Institute, Cary, North Carolina).

Results

Of 177 survivors of CHD surgery who met eligibility criteria, 143 survivors and their parents/guardians elected to participate. Survivors were an average of 12 ± 2.6 years of age, predominantly white, and slightly more likely to be male and from families above the US income median (Table I). Roughly one-third of survivors had single-ventricle heart lesions and, as expected, survivors of single-ventricle CHD had more surgeries and catheterizations, with fewer years since their last hospitalization ($P < .001$). About one-quarter of the sample had been diagnosed with a learning disability or had an individualized education program, with similar rates across single-ventricle and 2-ventricle CHD. Even so, survivors with single-ventricle CHD had higher rates of grade retention ($P < .001$) and missed more school in the past year ($P = .04$).

Aim 1: Complex CHD vs Normative Data on Executive Functioning, IQ, and School Performance

As predicted, survivors of complex CHD had significantly worse mean scores than norms on both behavior regulation and metacognitive aspects of executive functioning as well as IQ and school performance ($P \leq .001$; Table II). Unusually high rates of abnormal scores were present across all outcomes except BRIEF behavior regulation index, with one-quarter to one-half of survivors scoring in the worst 10% of norms on metacognition, IQ, and school performance ($P < .001$).

Of the 2 aspects of executive functioning, metacognition was more severely affected than behavior regulation ($t = 6.22$, $P < .0001$). Age correlated with poor metacognition ($r = .22$, $P = .007$) but not behavioral regulation ($r = .06$, nonsignificant). The prevalence of abnormal scores on the BRIEF metacognition index rose dramatically through adolescence (Figure).

Survivors with single-ventricle lesions were rated by their parents as having significantly lower school competency (mean = 39.9) and school QOL (mean = 56.7) than were survivors with 2-ventricle lesions (mean = 43.7, $P = .03$ and mean = 65.1, $P = .02$, respectively). Survivors with single-ventricle lesions themselves also reported significantly lower school QOL (mean = 60.5) than did survivors with 2-ventricle lesions (mean = 69.2, $P = .02$). In contrast, survivors with 2-ventricle lesions were rated as having poorer

Table I. Sample characteristics

	Total sample	Comparison of subsamples		
		1-ventricle	2-ventricle	*P* value
Sample size, n (%)	143	51 (36%)	92 (64%)	<.001
Demographics				
Male, n (%)	86 (60%)	31 (61%)	55 (60%)	.91
White, n (%)	119 (83%)	46 (90%)	73 (79%)	.10
Family Income below $51 000, n (%)	57 (43%)	20 (44%)	37 (42%)	.87
Age at time of study, mean (SD)	12.0 (2.6)	11.9 (2.6)	12.1 (2.6)	.69
Medical care utilization				
Number of cardiac surgeries, median (range)	2 (1-5)	3 (2-5)	1 (1-5)	<.001
Number of catheterizations/interventions, median (range)	2 (0-9)	3 (2-9)	1 (0-9)	<.001
Y since last hospitalization, median (range)	7.7 (0-16)	5.0 (0-15)	9.0 (0-16)	<.001
Patient education history				
Diagnosed with learning disability, n (%)	38 (27%)	14 (28%)	24 (26%)	.86
Have IEP, n (%)	38 (27%)	18 (35%)	20 (22%)	.08
History of grade retention, n (%)	30 (21%)	20 (39%)	10 (11%)	<.001
School wk missed in past year because of CHD, median (range)	0 (0-21)	0 (0-12)	0 (0-21)	.11
Total school wk missed in past y, median (range)	3 (0-60)	4 (0-23)	3 (0-60)	.04

IEP, Individualized Education Program.
Note: *P* value calculated based upon independent-sample *t* test for age, χ^2 test for dichotomous variables, and Wilcoxon rank sum test nonparametric tests.

Table II. Executive functioning, IQ, and school performance among survivors of complex CHD

Domain	Mean ± SD		Sample mean vs norms		Rates of abnormal scores in sample	
	CHD sample	Published norms	*t* test	*P*	% abnormal	*P*
Executive functioning*						
BRIEF behavior regulation index (T-score)	53.1 ± 11.3	50.0 ± 10.0	3.27	.001	13%	.42
BRIEF metacognition index (T-score)	56.6 ± 11.6	50.0 ± 10.0	6.84	<.001	24%	<.001
Intelligence						
Full-scale IQ (standard scores)	91.6 ± 16.3	100 ± 15.0	−6.12	<.001	22%	<.001
School functioning						
CBCL school competence (T-score)	42.3 ± 10.0	50.0 ± 10.0	−9.12	<.001	30%	<.001
Peds-QL school subscale – parent-report (raw)	62.1 ± 21.3	85.5 ± 17.6	−13.11	<.001	55%	<.001
Peds-QL school subscale – self-report (raw)	66.1 ± 21.4	78.6 ± 20.5	−6.94	<.001	25%	<.001

*Higher BRIEF scores indicate poorer functioning; higher IQ, CBCL, and Peds-QL scores indicate better functioning.
Source for Peds-QL norms: Varni et al 2001, p. 804-5.

behavioral regulation (mean = 55.1) than survivors with single-ventricle lesions (mean = 49.5, $P = .004$). There were no significant differences across heart lesion complexity for IQ or metacognition.

Aim 2: Executive Functioning Uniquely Predicts School Performance in CHD

Multivariable analysis assessed whether executive functioning was associated with school performance in the presence of demographic characteristics, heart lesion complexity, and IQ. Collectively, these variables predicted 37%-54% of the variance in parent-reported school competence and school QOL and in child-reported school QOL (Table III). The variance inflation scores were slightly above 1, indicating an acceptable level of collinearity in the models. Even after accounting for other predictors, poorer metacognition significantly predicted poorer parent-report school competence ($\beta = -.50$, $P < .001$) and school QOL ($\beta = -.55$, $P < .001$), and poorer child-reported school QOL ($\beta = -.37$, $P < .001$). Although both IQ and demographic factors were related to school performance, metacognition was the strongest single predictor, exceeding IQ and demographic factors. Indeed, neither behavior regulation nor heart lesion complexity predicted significant variance on any of the 3 school performance outcome measures in the presence of the other predictors. In other words, the effect of CHD complexity on school performance is not statistically significant when metacognition and IQ are in the models.

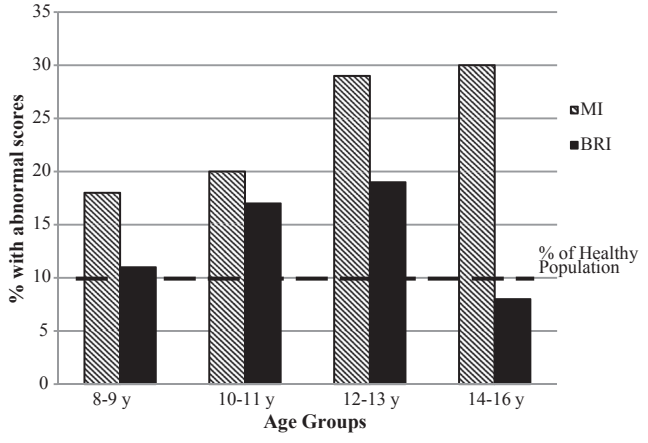

Figure. Executive functioning difficulties by age within complex CHD sample. *BRI*, behavioral regulation index; *MI*, metacognition index.

Discussion

Despite impressively improved survival rates for children with complex CHD, many survivors face significant neurodevelopmental morbidities that impact everyday life. The current study focused on potential deficits in executive functioning in everyday life, especially as it relates to school performance, and yielded 3 main findings. First, school-aged survivors of complex CHD have greater executive functioning difficulties in daily life than their heart-healthy peers. These findings are largely consistent with the few existing studies involving structured tests of executive functioning[8,19,20] and further demonstrate that pediatric survivors of complex CHD surgery evidence executive functioning deficits in real-world settings.[21] This distinction is important because results of office-based tests of executive functioning can be poor predictors of real-world functioning.[18] In real-world settings, the ability to regulate behaviors (eg, flexibly adapting to situations, inhibiting impulses) and to engage in metacognitive tasks (eg, planning, organization, follow-through) are critical to setting and achieving important life goals, including success at the primary "work" of childhood: functioning at school.

The second major finding was that deficits in metacognitive functioning were not only uniquely associated with school performance but more strongly predicted problems at school than IQ, race, sex, or heart lesion complexity. This finding highlights a previously underrecognized contributor to the poor scholastic functioning that was reflected in one-quarter to one-half our sample. Although parent report was used to measure both metacognition and

Table III. Multivariable analysis of child- and parent-reported school functioning

	School functioning					
	CBCL parent		Peds-QL parent		Peds-QL child	
Variables	b	β	b	β	b	β
Age	0.64	0.17*	0.33	0.04	0.56	0.07
Sex	1.36	0.07	7.56	0.17*	0.83	0.02
Race (White/nonwhite)	1.47	0.05	0.97	0.02	9.50	0.16†
Median family income	1.86	0.09	8.59	0.20*	10.12	0.24*
CHD complexity	2.23	0.11	4.49	0.10	5.56	0.12
Missed school wk	−0.81	−0.07	−5.36	−0.21*	−4.70	−0.19†
IQ	0.28	0.43‡	0.23	0.17†	0.16	0.12
BRIEF: metacognition	−0.42	−0.50‡	−1.01	−0.55‡	−0.67	−0.37‡
BRIEF: behavior regulation	0.14	0.17	0.23	0.12	0.20	0.11
Model statistics	$R^2 = 0.54$ (F = 17.96, $P < .001$)		$R^2 = 0.54$ (F = 18.29, $P < .001$)		$R^2 = 0.37$ (F = 9.73, $P < .001$)	

*$P < .01$.
†$P < .05$.
‡$P < .001$.

some aspects of school functioning, the link between the two cannot be attributed simply to reporter bias because metacognitive functioning was also a strong predictor of child-reported school performance, and parent report of behavior regulation predicted neither parent- nor child-reported school functioning. Although present findings replicated past work indicating a downward shift in overall IQ,[27,28] it appears that deficits in higher-level metacognitive processes may be even more important than IQ in influencing the overall success of children with complex CHD at school.

The final major finding was that, among executive functioning, aspects that relate to metacognitive skills (eg, planning, organization, follow-through) appear particularly salient to the outcomes of school-aged survivors of CHD. Behavior regulation was not a unique predictor of school performance and was much less likely than metacognitive functioning to be abnormal in our overall sample. Further, whereas behavior regulation did not vary by age, the risk of metacognitive difficulties increased with age. In this cross-sectional study, it is impossible to rule out cohort effects, but we posit that many children with CHD fail to keep up with the normal development of metacognitive skills. Executive functioning has a prolonged developmental course, paralleling the protracted maturation of the frontal lobes throughout childhood and adolescence.[29] As normative expectations increase developmentally, the effects of CHD on executive functioning may become more apparent. Cassidy et al[21] found that rates of executive functioning impairment were nearly twice as high for pre-adolescent and adolescent survivors than controls.

There were several limitations in the current study. First, survivors were recruited from a single region, which may impact the generalizability of the results. Study survivors may differ geographically, culturally, or ethnically from others with complex CHD. Second, study data were compared with normative standards, not a matched cohort that could afford more precise analysis. Even so, it is reassuring that the demographics of our sample (eg, slightly above median US income) might be expected to attenuate deficits, rather than accentuate them, and our sample scored similarly to other samples of children with complex CHD who had decreased cognitive functioning compared with both control groups and normative data.[30] Third, despite the large sample of survivors of CHD surgery, individual diagnostic groups were too small to allow for analyses beyond the rough 1- vs 2-ventricle grouping. Finally, the study relied on parent ratings as the sole measure of executive functioning. Office-based neuropsychological tests of executive functioning may fail to elicit deficits by virtue of their structured nature, but use of proxy-report also has inherent limitations (eg, respondent bias). Even remaining within the realm of proxy reporting, we recommend that future work collect teacher-report data to complement the caregiver perspective. Similarly, assessment of school functioning can be enhanced with objective scholastic measures and/or academic documentation.

Despite these limitations, current findings document the particular importance of metacognition as a key area of skill deficit in many survivors of complex CHD surgery that becomes even more prominent in late childhood. Knowledge of this vulnerability provides impetus for greater routine assessment of executive functioning for these pediatric survivors. This brief assessment, which may help to avert related scholastic struggles and underachievement, could be done by routinely asking about behavior regulation, planning, organization, and follow-through on tasks or systematically using questionnaires (such as the BRIEF) that offer the advantage of comparisons against age- and sex-linked norms. It may also keep executive functioning difficulties from being misattributed to personality or behavior problems (eg, laziness, boredom, stubbornness), which could delay or prevent implementation of appropriate supports. Our findings also suggest it is especially important to assess and monitor older children and adolescents, even if they have long been medically stable. Better early identification of potential executive impairments allows for opportunities for more specialized follow-up evaluation and interventions, typically by pediatric psychologists or neuropsychologists, in service of promoting

scholastic success in this at-risk population. Similar to other medically complex populations, as patients near young adulthood, consideration can also be given to the important role of executive functioning in transitioning planning and preparing for postsecondary opportunities.

In summary, executive functioning difficulties were significantly higher in pediatric survivors of complex CHD surgery, with metacognition emerging as a preeminent predictor of school functioning. These findings provide compelling evidence for further investigation, routine clinical assessment of metacognitive deficits, and interventions when needed, especially in older children and those who are struggling at school. ■

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