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Visuospatial processing in adolescents with critical congenital heart disease: Organization, integration, and implications for academic achievement

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Abstract

Among the most significant factors affecting quality of life in individuals with critical congenital heart disease (CCHD) are neurodevelopmental challenges, including deficits in visuospatial processing and academic achievement. Few studies have compared outcomes across CCHD subgroups, despite their significant differences in anatomy/physiology and medical/surgical courses. We compared visuospatial processing abilities (i.e., Developmental Scoring System, Rey Osterrieth Complex Figure; DSS-ROCF) across groups of adolescents with CCHD (d-transposition of the great arteries [TGA, $n = 139$], Tetralogy of Fallot [TOF, $n = 68$], single-ventricle cardiac anatomy requiring the Fontan operation [SVF, $n = 145$]) and a group of healthy referents (REF, $n = 111$), and examined the validity of visuospatial processing in predicting concurrent academic outcomes. The CCHD subgroups differed in Organization, ps < .001, Structural Accuracy, ps < .001, and Incidental Elements Accuracy scores, ps ≤ .008; post-hoc analyses showed that the SVF group tended to underperform other CCHD groups. With respect to academic skills, all CCHD groups had worse scores than the REF group, ps ≤ .007; CCHD groups were not different from each other, ps > .23. Regression results showed that DSS-ROCF Style rating (reflecting integration) accounted for a small yet statistically significant portion of unique variance in "assembled" academic outcomes, over and above the variance already accounted for by

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DSS-ROCF Organization, $p < .01$. Findings support the need for comprehensive neuropsychological assessment and monitoring of children and adolescents with CCHD, as well as targeted intervention for organization and integration deficits that may increase their risk for academic underachievement.

Keywords

Rey Osterrieth Complex Figure; Visual-spatial; Cardiac; Congenital Heart Defect; Adolescence

According to recent estimates, nearly two million people in the United States are living with congenital heart disease (CHD; Hoffman & Kaplan, 2002; Marelli et al., 2012; Reller, Strickland, Riehle-Colarusso, Mahle, & Correa, 2008); and, of these, approximately 25% are diagnosed with a critical CHD (CCHD) requiring intensive medical/surgical intervention(s) within the first year of life (Oster et al., 2013). As more children born with CCHD are surviving into adolescence and adulthood, it has become increasingly important to understand their neurodevelopmental trajectories. Although, as a group, children with CCHD tend to exhibit intellectual reasoning abilities in the low average to average range for age, they remain at risk for wide-ranging deficits in speech/language abilities, attention, executive function skills, self-regulation, fine/gross motor skills, academic achievement, and social cognition as they mature into school-age and adolescence (Bellinger & Newburger, 2010; Gaynor et al., 2009). Indeed, these neurodevelopmental and behavioral concerns are among the most significant factors affecting quality of life in those with CCHD, more significant even than delayed medical complications (Marino et al., 2012; Wernovsky, 2006).

Deficits in visuospatial processing have also been identified among children and adolescents with CCHD (e.g., Bellinger, Bernstein, Kirkwood, Rappaport, & Newburger, 2003); however, heterogeneity within samples and lack of consistency in the operationalization of visuospatial constructs and assessment methodologies used across studies make it difficult to draw firm conclusions about the impact of CCHD on the development of visuospatial skills. This study had two primary aims: 1) to assess several components of visuospatial processing, in a standardized fashion, across three groups of adolescents with CCHD [dextro-transposition of the great arteries (d-TGA), Tetralogy of Fallot (TOF), and single-ventricle cardiac anatomy requiring the Fontan procedure (SVF)], as well as a group of healthy referents (REF), and 2) to investigate the real-world functional relevance of visuospatial processing on concurrent academic achievement.

Visuospatial processing and the Rey-Osterrieth Complex Figure (ROCF).

Historically, visuospatial processing has been among the more difficult cognitive domains to define, in part because it is non-verbal and thus harder to characterize in words. It has been operationalized in numerous ways using many different measures. One of the most frequently used measures is the ROCF, which is a multi-faceted task requiring visuoperceptual, visuoconstructional, graphomotor, visual memory, and executive function skills to complete. Since its introduction by Andre Rey over 70 years ago, several scoring protocols have been developed for the figure, each emphasizing a different aspect of visuospatial processing ability (Davies, Field, Andersen, & Pestell, 2011). The

Developmental Scoring System (DSS), created by Bernstein and Waber (1996), is a developmentally-informed method of scoring the ROCF that allows for comparison of organizational precision and correctness (i.e., “Organization”), as well as the presence of figure elements (i.e., “Accuracy”). Accuracy is further divided into Structural Elements (i.e., those features associated with the organizational frame of the figure) and Incidental Elements (i.e., those features that are more peripheral details). Important for this study and discussed further below, the Developmental Scoring System, Rey Osterrieth Complex Figure (DSS-ROCF) also characterizes an individual’s style in approaching their drawings by rating the level of integration (or cohesion) in how the figure was reconstructed.

Visuospatial processing in children and adolescents with CCHD.

In a series of larger studies from which the data for this investigation were drawn, Bellinger and his colleagues (2003, 2011, 2014, 2015) demonstrated that children and adolescents with CCHD have difficulty managing complex visuospatial materials. At age 8 years, children with *dextro-transposition of the great arteries* (TGA) struggled to organize the DSS-ROCF. Among those in the lowest basal level on the DSS-ROCF (i.e., showing the poorest organization/precision in their reconstruction), poor performance was associated with deficits in math. Interestingly, this same cohort at 16 years old was indistinguishable from healthy referents on the DSS-ROCF. In contrast, adolescents with surgically-palliated *tetralogy of Fallot* (TOF) and those with *single-ventricle cardiac conditions requiring the Fontan procedure* (SVF) performed below expected levels on the DSS-ROCF, at least in terms of Organization and Accuracy. Unfortunately, none of these studies reported on *how* the adolescents approached the task, specifically, the level of integration with which they were able to perceive and manage the figure.

Despite significant differences in anatomy/physiology and medical/surgical courses across various forms of CCHD, few studies have attempted to compare neurodevelopmental outcomes across subgroups of CCHD.[1] In one study, Oates, Simpson, Cartmill, and Turnbull (1995) found no differences in group performance on the ROCF between cyanotic and acyanotic CHD groups; however, it is unclear what ROCF scoring system was used and, as such, what specific variables were included in the analysis. Moreover, relatively small sample sizes (*n*s = 30, 51, and 33, respectively) may have limited their power to find group

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[1] There are many subtypes of CCHD affecting the structure of the heart and/or the surrounding blood vessels, as well as the functional integrity of the cardiovascular system. Three subtypes of congenital heart defects in the current study include: TGA, TOF and SVF. All three are considered cyanotic conditions, meaning that the blood being circulated throughout the body is oxygen-deficient; all are considered CCHD, meaning that children are severely ill during the neonatal period and require surgical correction for survival (Hoffman & Kaplan, 2002; Miatton, De Wolf, Francois, Thiery, & Vingerhoets, 2006). For children born with *TGA*, the two main arteries carrying blood from the heart to the body are transposed; therefore, oxygenated blood continues to circulate to and from the lungs, while deoxygenated blood continues to circulate throughout the rest of the body (including the brain). Prior to surgery, survival is dependent on leakages (that allow some oxygenated blood to seep into the body through chambers in the heart) or catheterization until full surgical correction is possible, thus putting children in a state of suboptimal oxygenation until surgical correction is performed (arterial switch operation; “American Heart Association,” 2015). Children born with *TOF* have four co-occurring cardiac malformations: (1) ventricular septal defect, (2) pulmonary stenosis, (3) an overriding aorta, and (4) right ventricular hypertrophy. TOF is commonly associated with genetic conditions (e.g., 22q11.2 deletion syndrome) but can occur in isolation or in combination with other heart defects. TOF lesions result in some deoxygenated blood being pumped back into the body, thus reducing the overall concentration of oxygenated blood. These children require a single surgical correction or several staged procedures, depending on the severity of the cardiac lesions (“American Heart Association,” 2015). Children born with *single-ventricle lesions* have a rare disorder in which one of the lower chambers of the heart is malfunctioning or absent. Compared to other children with CCHD, these children are considered among the highest-risk and sustain a prolonged period of suboptimal cerebral oxygenation while completing staged surgical palliation (Fenton et al., 2007).

differences. In general, children with CCHD, compared to milder forms of cardiac disease, experience more significant neurodevelopmental impairment (Karsdorp, Everaerd, Kindt, & Mulder, 2007; Wernovsky, 2006), suggesting that differences in visuospatial skills may exist as a function of disease severity. It is important to study neurocognitive functioning across groups of different CCHD pathology, as general statements about "children with CHD" may not adequately capture differences across groups or provide guidance for clinicians working with individual children/adolescents.

Visuospatial organization, integration, and academic achievement:

In clinical settings, an individual's performance on the ROCF is often talked about as providing insight into that person's "cognitive style" (i.e., how he or she approaches and organizes complex, novel information). The concept of *integration* is frequently invoked. For the purposes of this discussion, we use *integration* to mean an individual's ability to appreciate connections and relationships between pieces of visuospatial information. Whereas some tend to approach materials in a detail-focused, part-oriented fashion, others take a more integrated, configurational, gestalt-oriented approach, making use of underlying organizational structures to frame and anchor their efforts. The DSS-ROCF is a process-oriented scoring framework that operationalizes cognitive style/integration in a developmentally-informed way and provides a standardized means of defining the level of integration (Bernstein & Waber, 1996), thus enabling the examination of the putative links between integration and real-world functional outcomes such as academic achievement.

Integration is considered essential for learning, particularly for academic tasks such as reading comprehension and applied mathematical problem solving, both of which require prioritizing and synthesizing multiple elements into a meaningful and coherent whole. These so-called "assembled" processing tasks may be contrasted with relatively more discrete, data-driven or "associative" processing tasks (i.e., based on the formation/activation of associations), such as single-word decoding and rote arithmetic calculation. These associative tasks should, in theory, rely less on integration and more on organization and detail-management (see Dennis, Landry, Barnes, & Fletcher, 2006 for further elaboration of this model in the spina bifida population).

Despite its frequent use in clinical and research neuropsychological batteries and its sensitivity to detecting central nervous system dysfunction in children, concurrent/predictive validity studies using the ROCF with children are limited. Davies et al. (2011) reported on four ROCF scoring methods and found that all were correlated with parent-report of cognitive and academic functioning (but not executive function skills). In adolescents after open-heart surgery, von Rhein and colleagues (2015) found that the ROCF was not correlated with other measures of cognitive functioning, suggesting that the task may be capturing a unique cognitive process. These authors also found that scoring systems based exclusively on quantitative aspects of ROCF performance (versus more process-oriented systems) were not sensitive to group differences.

Academic outcomes in CCHD.

For school-age children and adolescents, academic skills are an important marker of overall adjustment and can elucidate the functional impact of neurodevelopmental difficulties. Several studies have documented academic challenges in school-age children with CHD. In a large-scale follow-up study of children with a variety of complex cardiac pathologies (Shillingford et al., 2008), 18% of children had repeated a grade and 14–20% were rated by teachers as functioning “well below average.” A sizable portion of the sample (15%) was placed within a full-time special education classroom. Importantly, of the remaining group, 21% were getting reading/math support in the classroom and 13% were getting reading/math support outside the classroom, suggesting that the academic struggles in this group ranged from severe to mild, but were affecting about half of the sample. Shillingford et al. (2008) were unable to identify any significant associations between the need for remedial services and pre-, intra-, or post-operative variables; however, other investigations have shown that children with cyanotic CHD perform more poorly than those with acyanotic CHD on arithmetic, spelling, and reading tasks (Wray & Sensky, 2001).

Broadly-defined academic outcomes from the participants in this study have been reported previously. Math and reading composite scores show that participants with TGA score within the average range, yet significantly below expected population means (Bellinger et al., 2011). Participants with TOF showed slightly (although not significantly) lower performance than population means, with those adolescents with an identified genetic syndrome underperforming those without a genetic syndrome (Bellinger et al., 2014). Adolescents with SVF were also found to perform significantly below population means (Bellinger et al., 2015). Although these findings suggest that adolescents with CCHD are having more academic struggles than their peers, they do not address the particular aspects of reading and math that may be problematic, thus limiting their potential to inform intervention.

Current aims and hypotheses.

The current study had two primary aims: 1) to compare visuospatial processing abilities across three groups of adolescents with CCHD (TGA, TOF, SVF) and a group of healthy referents (REF), and 2) to examine the concurrent validity of visuospatial processing on academic outcomes. We expected that the SVF group would demonstrate more significant deficits in visuospatial processing than the other groups, given the severity of their cardiac disease. We also hypothesized that visuospatial organization and integration abilities would be differentially associated with academic outcomes. Specifically, while we expected *organization* to account for significant variance in both “associative” and “assembled” academic outcomes, we predicted that *integration* would account for unique variance in “assembled” (but not “associative”) academic outcomes.

Method

Data Collection

Data for this analysis were pooled from data collected during the course of three large-scale studies of neurodevelopmental outcomes in adolescents with CCHD (Bellinger et al., 2011, 2014, 2015). All three studies included careful medical inclusion and exclusion criteria, as well as extensive, age-appropriate neuropsychological assessments. Please see Cassidy, White, DeMaso, Newburger, and Bellinger (2015) for a detailed review of recruitment and data collection methodology.

Participants

Participant groups included children with d-transposition of the great arteries (TGA; Bellinger et al., 2011), Tetralogy of Fallot (TOF; Bellinger et al., 2014), single-ventricle cardiac anatomy requiring the Fontan operation (SVF; Bellinger et al., 2015), and healthy referents (REF).

Data for the *TGA group* were collected when the participants were 14–16 years old. These children underwent the arterial switch operation as infants and were randomly assigned to either cardiopulmonary bypass with predominant deep hypothermic circulatory arrest or low-flow bypass. Participants in the *TOF group* included 13–16 year olds who underwent surgical repair at least 6 months prior to assessment. The *SVF group* included participants between the ages of 10–19 years, who underwent surgical correction at least 6 months before assessment. The *REF group* included healthy adolescents ages 10–19 years who participated in the original TOF and SVF studies and were recruited to match admission criteria for the NIH MRI Study of Normal Brain Development (Waber et al., 2007).

Participants with identified genetic/syndromic conditions (e.g., 22q11.2 deletion syndrome) were excluded from analyses (TGA $n = 0$; TOF $n = 23$; SVF $n = 11$). The final analyses included four participant groups: TGA ($n = 139$), TOF ($n = 68$), SVF ($n = 145$), and REF ($n = 111$). Analyses were run with pair-wise exclusions to account for missing data [6 participants (1.3%) were unable to complete the DSS-ROCF; TGA $n = 1$; TOF $n = 3$; SVF $n = 1$; REF $n = 1$]. Table 1 presents patient demographics and comparability between groups.

Measures

Previous reports (Bellinger et al., 2011; 2014; 2015) have described overall DSS-ROCF and Wechsler Individual Achievement Test, Second Edition (WIAT-II; Math and Reading Composite scores) performance among CCHD and REF groups. This study provides a more detailed analysis of visuospatial processing and academic achievement outcomes, including previously-unpublished elements from the DSS-ROCF and individual WIAT-II subtest scores.

Rey-Osterrieth Complex Figure (ROCF).—The ROCF is a widely-used neuropsychological assessment tool used to assess visuospatial processing, visuo-motor functioning, and aspects of executive control (including organization and integration). Participants are asked to copy a complex figure while looking at the stimuli; they are asked to switch between different colored markers/pens in order to track their approach to the task. Participants are then asked to redraw the figure from memory immediately after the Copy trial (Immediate Recall trial) and following a 15–20 minute delay interval (Delayed Recall trial) using just one color/pen. Due to time constraints, the SVF group did not complete the Delayed Recall trial.

Figures were scored using the Developmental Scoring System (DSS-ROCF; Bernstein & Waber, 1996). The primary variables of interest for the Copy, Immediate Recall, and Delayed Recall trials included: Organization (DSS-ROCF Basal Level) and Accuracy (DSS-ROCF Structural and Incidental Elements); for both measures, higher scores reflect more accurate and complete drawings. Copy Trial Style ratings were used to characterize the level of integration; from least to most integrated, Copy trial reproductions were rated as: Part-Oriented, Intermediate (OC/IP: Outer Configurational/Inner Part; OP/IC: Outer Part/Inner Configurational), or Configurational. Style ratings are referenced to the Basal Level; therefore, Basal Level was included as a covariate in analyses of integration.

Wechsler Individual Achievement Test, Second Edition (WIAT-II).—WIAT-II age-normed standard scores were used to measure academic skills in reading and mathematics. WIAT-II outcomes were divided into two groups: (1) discrete/associative tasks: Word Reading, Pseudoword Decoding, and Numerical Operations subtests and (2) assembled tasks: Reading Comprehension and Mathematics Reasoning subtests.

Data Analytic Approach

Patient demographics and clinical characteristics were summarized using descriptive statistics and compared across groups. Socioeconomic status (SES), gestational age, birthweight, and age at assessment were compared across groups using Analysis of Variance (ANOVA); if a significant group effect was found, pairwise comparisons among groups were adjusted for multiple comparisons using Tukey's honest significant difference (HSD) test. Age at first operation and total cardiac operations were skewed; therefore, CCHD group comparisons for these variables were evaluated using the Kruskal-Wallis test (nonparametric ANOVA). If a significant group effect was found, the Dwass, Steel, Critchlow and Fligner multiple comparison procedure was used to identify significant pairwise differences (Crichtlow & Fligner, 1991; Dwass, 1960; Steel, 1960). Gender and race were compared across groups using Fisher's exact test. If a significant group effect was found, all pairwise comparisons were evaluated and a false discovery rate (FDR) procedure (Benjamini & Hochberg, 1995) was subsequently performed to identify significant group differences.

The Copy, Immediate Recall, and Delayed Recall trial Organization results were compared across groups using ordinal logistic regression. The Copy, Immediate Recall, and Delayed Recall trial results for the Structural Elements Accuracy and Incidental Elements Accuracy measures were dichotomized to account for highly skewed performance ("perfect/less than perfect" Structural Elements and ">75th percentile/≤75th percentile" Incidental Elements) and analyzed using logistic regression. For each outcome above, the model included a main effect for group, and a forward selection procedure was used to identify additional significant covariates to include in the model; the candidate covariates included age, race, and SES. Pairwise group comparisons were adjusted using Tukey's HSD test.

The frequency and percent of participants in each Style rating category were calculated by group. The association between Style rating and group was assessed using ordinal logistic regression, and pairwise group comparisons were adjusted using Tukey's HSD test. For each of the ordinal regression findings described above, the "odds" presented refer to the odds of receiving a lower/worse category rating or score.

The WIAT-II subtest scores were compared across groups using Analysis of Covariance (ANCOVA). For each subtest, the model included a main effect for group, and a forward selection procedure was used to identify additional significant covariates to include in the model; the candidate covariates included age, race, SES, gender, birth weight, and gestational age. Pairwise comparisons among groups were adjusted for multiple comparisons using Tukey's HSD test.

Composite variables were created by averaging across discrete ("associative") and "assembled" WIAT-II tasks, respectively. Separate hierarchical linear regressions (controlling for age, SES, and DSS-ROCF Copy Basal Level) were used to test associations between visuospatial processing (Integration and Organization) abilities and discrete/associative and assembled academic outcomes, respectively. Control variables were entered at step 1. CCHD group status (CCHD vs. REF) was entered at step 2. DSS-ROCF Copy Organization score was entered at step 3. DSS-ROCF Copy Style rating (part-oriented vs. intermediate vs. configurational) was entered at step 4.

Analyses were conducted using SAS version 9.3 or SPSS version 23. All tests were 2-sided at the 0.05 significance level.

Results

Organization.

Overall, there were significant group differences in level of organization (i.e., DSS-ROCF Basal Level) for the Copy, X^2 (3, N = 455) = 44.12, $p < .001$, and Immediate Recall trials, X^2 (3, N = 455) = 24.84, $p < .001$. Group performance was equivalent on the Delayed Recall trial, X^2 (2, N = 261) = 3.42, $p = .18$. Post-hoc analyses revealed that Copy Organization scores were lower in the SVF group than all other groups, *p*s ≤ .001. No differences emerged in Copy Organization between TGA, TOF, and REF groups. On the Immediate Recall trial, the SVF group was more likely than REF group to score lower on Organization, $p < .001$. Post-hoc analyses revealed that Immediate Recall Organization scores were lower in the SVF group than the TGA group as well, $p < .05$.On the Delayed Recall trial, no differences were found between TGA, TOF, or REF groups (note: the SVF group did not participate in this trial, as described above). Table 2 presents group differences in the odds of receiving a lower Basal Level score.

Accuracy: Structural Elements

Overall, there were significant group differences in Structural Accuracy scores for the Copy, X^2 (5, N = 455) = 51.61, $p < .001$, Immediate Recall, X^2 (5, N = 456) = 75.27, $p < .001$, and Delayed Recall trials, X^2 (4, N = 261) = 39.75, $p < .001$. On the Copy Trial, relative to the REF group, the odds of scoring less-than-perfect on Structural Elements were approximately 6 times higher for TOF($p = .01$), and SVF groups ($p = .002$), but were not significantly higher for TGA ($p = .28$). On the Immediate Recall trial, SVF ($p < .0001$), and TOF groups ($p = .01$), were more likely than the REF group to achieve less-than-perfect Structural Elements scores. On the Delayed Recall trial, the TOF group was more likely than the REF group to achieve less-than-perfect Structural Elements scores ($p < .001$). Table 2 presents group differences in the odds of receiving a lower Structural Elements Accuracy score.

Accuracy: Incidental Elements.

Overall, there were significant group differences in Incidental Elements Accuracy scores for the Copy, X^2 (5, N = 455) = 46.29, $p < .001$, and Immediate Recall trials, X^2 (5, N = 455) = 15.67, $p = .008$. No significant group differences in Incidental Elements were found on the Delayed Recall trial, X^2 (4, N = 261) = 4.41, $p = .35$. On the Copy Trial, odds of scoring ≤75th percentile on Incidental Elements were approximately 3 times higher for TOF ($p = .02$) and SVF ($p < .001$) versus the REF group. On the Immediate Recall trial, the SVF group was approximately 3 times more likely to score ≤ 75th percentile for Incidental Elements than the REF group ($p = .04$). Table 2 presents group differences in odds of receiving a lower Incidental Elements Accuracy score.

Post-hoc analyses revealed that Immediate Recall Structural and Incidental Elements scores were lower in the SVF group than the TGA group (ps < .05).

Integration.

Table 3 presents percentages of each Style Rating category by group. Results of ordinal logistic regression revealed that the SVF group had 2.5 times the odds of having a lower Style rating than the TGA group ($p < .001$) and 1.9 times the odds of having a lower Style rating than the REF group ($p = .05$). Other comparisons were not significant (ps > .26).

Academic Achievement.

Table 4 presents group performance on academic achievement subtests. Controlling for significant covariates, there were significant group differences in academic performance across all tasks: Word Reading, $F(3, 458) = 17.7$, $p < .001$, Pseudoword Decoding, $F(3, 457) = 12.6$, $p < .001$, Numerical Operations, $F(3, 459) = 18.5$, $p < .001$, Reading Comprehension, $F(3, 457) = 14.1$, $p < .001$, Mathematics Reasoning, $F(3, 458) = 17.7$, $p < .001$. The REF group outperformed all CCHD groups, ps ≤ .007. The CCHD groups were not significantly different from each other, ps > .23.

Predictive value of Integration and Organization for Academic Outcomes.

Regression results are presented in Table 5. We tested separate hierarchical regression models for “discrete/associative” and “assembled” academic outcomes. In the “discrete/

associative" model, all three covariates (age, SES, DSS-ROCF Copy trial Basal Level), CCHD group, and DSS-ROCF Organization scores accounted for significant unique variance in discrete/associative academic skills over and above the variables entered earlier in the model; DSS-ROCF Style rating (i.e., "integration") did not account for unique variance over and above the other variables in the model.

In the "assembled" model, once again all three covariates (age, SES, DSS-ROCF Copy trial Basal Level), CCHD group, and DSS-ROCF Organization scores accounted for significant unique variance in assembled academic skills over and above the variables entered earlier in the model. However, in contrast to the discrete/associative model and consistent with our hypothesis, DSS-ROCF Style rating accounted for a small yet statistically significant portion of unique variance in "assembled" academic outcomes, over and above the variance already accounted for by DSS-ROCF Organization and the other variables in the model.

Discussion

Consistent with prior research, our findings show that adolescents with a history of CCHD are at risk for deficits in visuospatial processing. More specifically, findings highlight that patterns of visuospatial processing differ by type of CCHD. Furthermore, we explored the real-world concurrent validity of visuospatial processing difficulties, with findings implicating both organization and integration as important for academic skill development. As we hypothesized, integration was differentially and uniquely implicated in "assembled" academic outcomes.

Visuospatial processing and CCHD.

Adolescents with single-ventricle cardiac conditions who underwent the Fontan procedure (SVF) had the highest risk for visuospatial organization deficits, performing significantly worse than those with other forms of CCHD (i.e., TGA and TOF) and healthy referents. Beyond organization, Structural Elements Accuracy scores (reflecting the presence of elements and prioritization of the structural frame of the image) were particularly sensitive to cardiac status with a nearly 6 times greater odds of impairment among adolescents in both the TOF and SVF groups versus healthy referents (REF). Immediate visuospatial memory also differed by cardiac condition, with both TOF and SVF groups scoring below referents and SVF participants scoring below TGA participants. Taken together, these findings suggest that when tasked with managing complex visuospatial materials, many adolescents with CCHD do not effectively prioritize key structural/organizational features, focusing instead on incidental details that, while salient, may nonetheless provide a less robust framework upon which to encode and recall materials at a later time.

Style Ratings were used to capture participants' approach to the ROCF using a standardized rating system. Participants in the SVF group were more likely than the TGA and referent groups to approach the task in a poorly-integrated, part-oriented manner. Many rating systems for the ROCF were originally designed for adults, making extension into pediatric populations more challenging, and do not include a standardized mechanism for assessing an individual's process or approach to the task. The current findings support previous assertions that utilizing a standardized approach to measuring visuospatial processing in research studies, as well as clinical assessment, may aid in elucidating group differences not otherwise detected by systems focused predominately on presence/absence of features and precision (von Rhein et al., 2015).

The factors affecting brain and cognitive development in children and adolescents with CCHD are varied and include pre-, intra- and post-operative influences (Wernovsky, 2006). Neurobehavioral outcomes are likely related to a complex interaction of patient specific factors (e.g., genetic susceptibility) and environmental factors (e.g., surgery, SES) (Gaynor et al., 2007), suggesting that influences other than intraoperative management may be important determinants of neurobehavioral outcomes for many children with CCHD as well. This complexity in central nervous system involvement in CCHD precludes finding a single predictive variable to explain CCHD group differences. However, when comparing across subtypes of CCHD, possible explanations for the increased visuospatial processing difficulties demonstrated in the SVF group could be related to disease burden; as described above, these children are considered among the highest risk of those with CCHD and many endure a prolonged period of suboptimal cerebral oxygenation while undergoing staged surgical palliation (Fenton, Lessman, Glogowski, Fogg, & Duncan, 2007). Even prior to surgical intervention, some reports have documented evidence of brain injury in up to 36% children with single-ventricle physiology, with findings suggesting that reduced fetal cerebral blood flow is linked to delayed brain development in utero (Sethi et al., 2013). Furthermore, the SVF group as a whole tends to require more surgical procedures (see Table 1) and, relatedly, are exposed to more general anesthesia than other CCHD groups – both of which pose significant risk for the developing brain (Jevtovic-Todorovic et al., 2013). Interestingly, studies with school-age children awaiting cardiac surgery show evidence of visuospatial difficulties prior to cardiac surgery (in particular, immediate recall of Structural Elements compared to healthy controls), with a similar pattern in those patients awaiting their first versus follow-up surgery (van der Rijken et al., 2010); these findings highlight that visuospatial difficulties in CCHD are not exclusively related to surgical factors/ complications and are more likely related to a composite (and potentially cumulative) set of factors related to disease burden and environmental modifiers (e.g., early intervention services).

Numerous hypotheses have been put forth to explain why visuospatial processing seems to be more commonly affected among medically-complex populations compared to other cognitive processes. Considering CCHD specifically, some have suggested that early experiential factors such as hospitalizations and multiple operations may interfere with normal motor/spatial development (Karsdorp et al., 2007). Neuroanatomical “crowding” hypotheses have also been posited, whereby language is thought to have privileged status over visuospatial processing, and thus tends to be more robust in the face of early injury (due to its propensity to overtake brain areas that would otherwise have been devoted to visuospatial skills). There is also the possibility, as Stiles and her colleagues highlighted in their seminal perinatal stroke studies, that evolutionarily older and relatively more circumscribed visuospatial regions may be endowed with less plasticity than the evolutionarily younger, more widely distributed, and thus more resilient language system (Stiles, Reilly, Levine, Trauner, & Nass, 2012).

Several authors have emphasized that, given the normal IQs characteristic of this population, comprehensive neuropsychological assessment is necessary in order to identify neurocognitive areas of need (e.g., Daliento, Mapelli, & Volpe, 2006; Marino et al., 2012). With the literature documenting the significant morbidity of neurocognitive, social, and academic capacities in this population, neuropsychologists are increasingly called upon to evaluate the breadth and severity of impact of CCHD on developmental progress (Bellinger & Newburger, 2010). Our findings highlight that comprehensive assessment of visuospatial processing (with both quantitative and qualitative observation) is important for children with CCHD, in addition to evaluation of more fundamental visuoperceptual skills.

Associations between visuospatial processing and academic achievement.

Finally, we examined the validity of the DSS-ROCF in predicting basic and higher-order academic competencies. Consistent with our hypotheses, whereas the ability to effectively organize complex visuospatial materials was associated broadly with academics (both “discrete/associative” and “assembled” skills), the degree of integration applied in managing those materials was uniquely associated with higher-order (“assembled”) academic skills.

During the early school years, the educational system (at least in the United States) tends to emphasize memorization of the “building blocks” – outfitting young learners with the discrete/associative processing capacities they need to begin understanding symbols, decoding words, and working with numbers. Over time, curricular demands become increasingly integrative – expecting students to not only acquire new discrete skills, but also to link new skills with old in an ever-expanding knowledge network that will ultimately permit them to reach higher conceptual and computational heights. Children with CCHD may struggle with learning from very early on, or perhaps not until latent integration vulnerabilities become manifest in the shift from “learning to read” (i.e., “discrete/associative processing”) to “reading to learn” (i.e., “assembled processing”) that marks the transition from the early-elementary to later-elementary/early-middle-school years. The DSS-ROCF, as a means of operationalizing and measuring integration in children and adolescents with CCHD, may prove useful in recognizing risk for current and future academic difficulties, and informing recommendations for clinical management. For example, appreciating that children with SVF may tend to adopt a more part-oriented approach to managing complex information, a neuropsychologist might suggest that teachers help these children to appreciate links between information; by making underlying connections and links between information more explicit, these students may be more likely to appreciate (and prioritize) the structure or narrative holding information together, in turn making them more successful readers and problem-solvers.

Previous authors have emphasized that neurodevelopmental sequelae of CCHD can play an important role in limiting educational, occupational, financial, and quality of life attainment for survivors (Marino et al., 2012). Our findings are in line with this statement, showing that fundamental visuospatial processing is linked to educational outcomes. Marino and colleagues (2012) emphasized in their statement that children with CCHD require close monitoring, assessment, and intervention to best manage the increased risk for atypical development, with direct referral to developmental specialists appropriate in some cases.

Although the concurrent nature of the current results do not imply causality, if integration is, in fact, a primary contributing factor to academic struggles, recommendations and interventions targeted at increasing integration ability may lead to downstream positive effects on the emergence of higher-level, “assembled” academic skills. Findings support the need for careful neuropsychological assessment and monitoring of children and adolescents with CCHD, as well as targeted intervention and remediation of organization and integration deficits that may increase their risk for academic underachievement.

Limitations.

This study has some limitations that are important to discuss. First, the current analyses excluded children with known genetic syndromes. This exclusion criterion served to clarify results related to CCHD history, but may limit generalizability, for example, by underestimating group differences among children who are more severely affected (e.g., Bellinger, 2014). Also notable, not all of the participants in the study groups underwent genetic testing; therefore, there may be some children with unidentified genetic conditions who were not excluded. Second, combining three separate large-scale studies provided the power to detect group differences amongst children with different subtypes of CCHD; however, subtle methodological differences between the three studies did not allow for direct comparison in some instances. For example, we were unable to directly compare or control for intellectual reasoning due to different tools used in each study. The lack of ROCF Delayed Recall data in the SVF group is also notable.

Future Directions.

In future studies, it will be important to determine the causal relationship between visuospatial difficulties and academic outcomes, as well as to evaluate the effectiveness of associated recommendations for educational success. Adopting a standardized marker of disease severity and standardized assessment tools (e.g., ROCF-DSS) would allow for more reliable comparison across studies. These methodological considerations would help to enrich a literature that has historically been limited by small sample sizes. Furthermore, investigation of potential prenatal, medical, surgical and/or developmental/experiential factors that may contribute to CCHD-group differences in visuospatial processing would also help to identify children most at risk for difficulties early in development, in order to provide needed educational and other support services to mitigate the impact of this domain of neurocognitive weakness.

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References

American Heart Association. (2015). Retrieved from http://www.heart.org/HEARTORG/

Bellinger DC, Bernstein JH, Kirkwood MW, Rappaport LA, & Newburger JW (2003). Visual-spatial skills in children after open-heart surgery. Journal of Developmental and Behavioral Pediatrics, 24(3), 169–179. [PubMed: 12806229]

Bellinger DC, & Newburger JW (2010). Neuropsychological, psychosocial, and quality-of-life outcomes in children and adolescents with congenital heart disease. Progress in Pediatric Cardiology, 29(2), 87–92. 10.1016/j.ppedcard.2010.06.007

Bellinger DC, Watson CG, Rivkin MJ, Robertson RL, Roberts AE, Stopp C, … Newburger, J. W (2015). Neuropsychological status and structural brain imaging in adolescents with single ventricle who underwent the Fontan procedure. Journal of the American Heart Association, 4(12), e002302. [PubMed: 26667085]

Bellinger DC, Wypij D, Rivkin MJ, Demaso DR, Robertson RL, Dunbar-Masterson C, … Newburger, J. W (2011). Adolescents with d-transposition of the great arteries corrected with the arterial switch procedure: Neuropsychological assessment and structural brain imaging. Circulation, 124(12), 1361–1369. 10.1161/CIRCULATIONAHA.111.026963 [PubMed: 21875911]

Bellinger D, Rivkin M, DeMaso D, Robertson R, Stopp C, Dunbar-Masterson C, … Newburger J (2014). Adolescents with tetralogy of Fallot: Neuropsychological assessment and structural brain imaging. Cardiology in the Young, 25(2), 338–47. [PubMed: 24512980]

Benjamini Y, & Hochberg Y (1995). Controlling the false discovery rate: A practical and powerful approach to multiple testing. *Journal of the Royal Statistical Society*, *Series B*(Methodological), 289–300.

Bernstein JH, & Waber DP (1996). Developmental Scoring System for the Rey-Osterrieth Complex Figure (DSS-ROCF). Odessa, FL: Psychological Assessment Resources, Inc.

Cassidy AR, White MT, DeMaso DR, Newburger JW, & Bellinger DC (2015). Executive function in children and adolescents with critical congenital heart disease. Journal of the International Neuropsychological Society, 20, 34–49.

Crichtlow DE, & Fligner MA (1991). On distribution-free multiple comparisons in the one-way analysis of variance. Communications in Statistics-Theory and Methods, 20(1), 127–139.

Daliento L, Mapelli D, & Volpe B (2006). Measurement of cognitive outcome and quality of life in congenital heart disease. Heart, 92, 569–574. [PubMed: 16537785]

Davies SR, Field ARJ, Andersen T, & Pestell C (2011). The ecological validity of the Rey-Osterrieth Complex Figure: Predicting everyday problems in children with neuropsychological disorders. Journal of Clinical and Experimental Neuropsychology, 33(7), 820–831. [PubMed: 21957867]

Dennis M, Landry SH, Barnes M, & Fletcher JM (2006). A model of neurocognitive function in spina bifida over the life span. Journal of the International Neuropsychological Society, 12, 285–296. [PubMed: 16573862]

Dwass M (1960). Some k-sample rank-order tests. Contributions to Probability and Statistics, 198–202.

Fenton KN, Lessman K, Glogowski K, Fogg S, & Duncan KF (2007). Cerebral oxygen saturation does not normalize until after stage 2 single ventricle palliation. The Annals of Thoracic Surgery, 83(4), 1431–6. [PubMed: 17383352]

Gaynor JW, Nord AS, Wernovsky G, Bernbaum J, Solot CB, Burnham N, … Gerdes M (2009). Apolipoprotein E genotype modifies the risk of behavior problems after infant cardiac surgery. Pediatrics, 124(1), 241–250. 10.1542/peds.2008-2281.Apolipoprotein [PubMed: 19564306]

Gaynor JW, Wernovsky G, Jarvik GP, Bernbaum J, Gerdes M, Zackai E, … Spray TL (2007). Patient characteristics are important determinants of neurodevelopmental outcome at one year of age after neonatal and infant cardiac surgery. The Journal of Thoracic and Cardiovascular Surgery, 133(5), 1344–1353. 10.1016/j.jtcvs.2006.10.087 [PubMed: 17467455]

Hoffman JI, & Kaplan S (2002). The incidence of congenital heart disease. Journal of the American College of Cardiology, 39(12), 1890–1900. [PubMed: 12084585]

Hollingshead, A. B. (1975). Four factor index of social status.

Jevtovic-Todorovic V, Absalom AR, Blomgren K, Brambrink A, Crosby G, Culley DJ, … Hemmings HC, Jr (2013). Anaesthetic neurotoxicity and neuroplasticity: An expert group report and statement based on the BJA Salzburg Seminar. British Journal of Anaesthesia, 111(2), 143–151. [PubMed: 23722106]

Karsdorp PA, Everaerd W, Kindt M, & Mulder BJM (2007). Psychological and cognitive functioning in children and adolescents with congenital heart disease: A meta-analysis. Journal of Pediatric Psychology, 32(5), 527–541. [PubMed: 17182669]

Marelli A, Gilboa S, Devine O, Kucik J, Oster M, Riehle-Colarusso T, … Jenkins K (2012). Estimating the congenital heart disease population in the united states in 2010-what are the numbers? Journal of the American College of Cardiology, 59(13s1), E787–E787.

Marino BS, Lipkin PH, Newburger JW, Peacock G, Gerdes M, Gaynor JW, … Mahle WT (2012). Neurodevelopmental outcomes in children with congenital heart disease: Evaluation and management: A scientific statement from the American Heart Association. Circulation, 126(9), 1143–72. 10.1161/CIR.0b013e318265ee8a [PubMed: 22851541]

Miatton MA, De Wolf D, Francois K, Thiery E, & Vingerhoets G (2006). Neurocognitive consequences of surgically corrected congenital heart defects: a review. Neuropsychology Review, 16(2), 65–85. [PubMed: 16960756]

Oates RK, Simpson JM, Cartmill TB, & Turnbull JA (1995). Intellectual function and age of repair in cyanotic congenital heart disease. Archives of Disease in Childhood, 72(4), 298–301. [PubMed: 7763059]

Oster ME, Lee KA, Honein MA, Riehle-Colarusso T, Shin M, & Correa A (2013). Temporal trends in survival among infants with critical congenital heart defects. Pediatrics, 131(5), e1502–e1508. 10.1542/peds.2012-3435 [PubMed: 23610203]

Reller MD, Strickland MJ, Riehle-Colarusso T, Mahle WT, & Correa A (2008). Prevalence of congenital heart defects in metropolitan Atlanta, 1998–2005. The Journal of Pediatrics, 153(6), 807–813. [PubMed: 18657826]

Sethi V, Tabbutt S, Dimitropoulos A, Harris KC, Chau V, Poskitt K, … McQuillen PS (2013). Single-ventricle anatomy predicts delayed microstructural brain development. Pediatric Research, 73(5), 661–667. [PubMed: 23407116]

Shillingford AJ, Glanzman MM, Ittenbach RF, Clancy RR, Gaynor JW, & Wernovsky G (2008). Inattention, hyperactivity, and school performance in a population of school-age children with complex congenital heart disease. Pediatrics, 121(4), e759–67. 10.1542/peds.2007-1066 [PubMed: 18381503]

Steel RG (1960). A rank sum test for comparing all pairs of treatments. Technometrics, 2(2), 197–207.

Stiles J, Reilly JS, Levine SC, Trauner DA, & Nass R (2012). Neural plasticity and cognitive development: Insights from children with perinatal brain injury. Oxford University Press.

van der Rijken R, Hulstijn-Dirkmaat G, Kraaimaat F, Nabuurs-Kohrman L, Daniels O, & Maassen B (2010). Evidence of impaired neurocognitive functioning in school-age children awaiting cardiac surgery. Developmental Medicine and Child Neurology, 52, 552–558. [PubMed: 20002112]

von Rhein M, Kugler J, Liamlahi R, Knirsch W, Latal B, & Kaufmann L (2015). Persistence of visuo-constructional and executive deficits in adolescents after open-heart surgery. Research in Developmental Disabilities, 36, 303–310.

Waber DP, & Bernstein JH (1995). Performance of learning-disabled and non-learning-disabled children on the Rey-Osterrieth Complex Figure: Validation of the developmental scoring system. Developmental Neuropsychology, 11(2), 237–252.

Waber DP, De MC, Forbes PW, Almli CR, Botteron KN, Leonard G, … Rumsey J (2007). The NIH MRI study of normal brain development: performance of a population based sample of healthy children aged 6 to 18 years on a neuropsychological battery. Journal of the International Neuropsychological Society, 13(5), 729–746. 10.1017/S1355617707070841 [PubMed: 17511896]

Wernovsky G (2006). Current insights regarding neurological and developmental abnormalities in children and young adults with complex congenital cardiac disease. Cardiology in the Young, 16(1), 92–104. 10.1017/S1047951105002398

Wray J, & Sensky T (2001). Congenital heart disease and cardiac surgery in childhood: Effects on cognitive function and academic ability. Heart, 85, 687–691. [PubMed: 11359753]

Table 1

Participant Demographics and Comparability Across Groups

	TGA	TOF	SVF	REF	*p*	Pairwise Comparisons (*p* < .05)
	(*n* = 138–139)	**(*n* = 62–68)**	**(*n* = 141–145)**	**(*n* = 101–111)**		
Family SES[a]	45.8 (12.2)	48.7 (12.0)	49.5 (13.3)	53.1 (9.6)	< .001	SVF, REF > TGA
Gestational age (*weeks*)	39.8 (1.2)	39.2 (2.5)	39.0 (2.2)	39.6 (1.3)	.003	TGA > SVF
Birth weight (*kg*)	3.6 (0.4)	3.2 (0.7)	3.3 (0.6)	3.5 (0.6)	< .001	TGA > TOF, SVF REF > TOF
Male, *n* (%)	106 (76.3%)	38 (55.9%)	90 (62.1%)	59 (53.2%)	< .001	TGA > TOF, SVF, REF
Race/Ethnicity, *n* (%)[b]					.06	
White/Caucasian/Non-Hispanic	126 (90.7%)	59 (86.8%)	118 (81.4%)	89 (80.2%)		
White/Hispanic	5 (3.6%)	5 (7.4%)	16 (11.0%)	3 (2.7%)		
Asian	2 (1.4%)	0	4 (2.8%)	5 (4.5%)		
Black	2 (1.4%)	2 (2.9%)	5 (3.5%)	9 (8.1%)		
Other	4 (2.9%)	2 (2.9%)	2 (1.4%)	5 (4.5%)		
Age at assessment (*years*)	16.1 (0.5)	14.7 (1.2)	14.4 (2.9)	15.3 (1.8)	< .001	TGA > TOF, SVF, REF REF > SVF
Age at first operation (*days*), *Median (IQR)*	6.0 (4.0, 9.0)	113.0 (55.5, 209.0)	6.0 (3.0, 21.0)	--		TOF > TGA, SVF
Total cardiac operations), *Median (min, max)*	1 (1, 4)	2 (1, 7)	3 (1, 5)	--		

Note: Data are presented as *mean* (*standard deviation*) unless otherwise noted. SES, gestational age, birthweight, and age at assessment were compared among groups using ANOVA. Gender and race were compared among groups using Fisher’s exact test. Age at first operation and total cardiac operations were compared among groups using the Kruskal-Wallis test. Pairwise tests were adjusted for multiple comparisons.

[a] Hollingshead, A. B. (1975). Four-factor index of social status. Unpublished manuscript, Yale University, New Haven, CT.

[b] Categorized as non-Hispanic White versus other for significance testing.

Table 2

Odds of receiving a lower Basal Level/Accuracy score, compared to the healthy referent (REF) group

	TGA			TOF			SVF			CCHD-Group Pairwise Comparisons ($p < .05$)
	p	*OR*	*95% CI*	*p*	*OR*	*95% CI*	*p*	*OR*	*95% CI*	
Organization										
Copy	.99	1.1	.57 – 2.0	.72	1.3	.65 – 2.8	**< .0001**	**3.7**	**2.0 – 6.9**	TGA, TOF < SVF
Immediate Recall	.53	1.4	.75 – 2.6	.09	1.9	.93 – 4.0	**< .0001**	**3.0**	**1.6 – 5.5**	TGA < SVF
Delayed Recall	.83	.85	.44 – 1.7	.15	1.8	.85 – 3.9	--	--	--	--
Accuracy: Structural										
Copy	.28	2.7	.64 – 11.8	**.01**	**5.6**	**1.3 – 24.5**	**.002**	**6.4**	**1.7 – 24.4**	--
Immediate Recall	.23	1.7	.83 – 3.5	**.01**	**2.8**	**1.2 – 6.7**	**< .0001**	**5.8**	**2.7 – 12.4**	TGA < SVF
Delayed Recall	.09	2.1	.92 – 5.0	**<.001**	**4.4**	**1.7 – 11.3**	--	--	--	--
Accuracy: Incidental										
Copy	.08	2.3	.94 – 5.5	**.02**	**2.9**	**1.1 – 7.6**	**< .001**	**3.5**	**1.5 – 7.9**	--
Immediate Recall	.92	.82	.38 – 1.8	1.0	1.0	.38 – 2.7	**.04**	**2.6**	**1.0 – 6.6**	TGA < SVF
Delayed Recall	.95	.89	.35 – 2.3	.98	1.1	.36 – 3.2	--	--	--	--

Note: Organization = ROCF-DSS Basal Level; Accuracy: Structural = ROCF-DSS Structural Elements dichotomized score; Accuracy: Incidental = ROCF-DSS Incidental Elements dichotomized score; bolded entries reflect significant findings at the $p \leq .05$. Pairwise tests were adjusted for multiple comparisons.

Table 3

Percentage of Style Rating by group

	TGA	TOF	SVF	REF
Part-Oriented	35 (25.4%)	22 (33.8%)	67 (46.9%)	34 (31.2%)
Intermediate: OC/IP	65 (47.1%)	31 (47.7%)	58 (40.6%)	49 (45.0%)
Intermediate: OP/IC	10 (7.2%)	2 (3.1%)	3 (2.1%)	8 (7.3%)
Configurational	28 (20.3%)	10 (15.4%)	15 (10.5%)	18 (16.5%)

Note: Data presented as *n* (percentage).

Results of ordinal logistic regression revealed that the SVF group had 2.5 times the odds of having a lower Style rating than the TGA group ($p < .001$) and 1.9 times the odds of having a lower Style rating than the REF group ($p = .05$); other comparisons were not significant (*p*s > .26). Pairwise tests were adjusted for multiple comparisons.

Table 4

WIAT-II Subtest and Composite Scores by Group

	TGA	**TOF**	**SVF**	**REF**	**Pairwise Comparisons ($p < .05$)**
“Discrete/Associative” Composite	97.0 (14.5)	97.3 (17.9)	94.5 (15.8)	108.0 (8.9)	REF > TGA, TOF, SVF
Word Reading	97.6 (15.3)	98.5 (17.7)	95.3 (17.6)	108.7 (9.0)	REF > TGA, TOF, SVF
Pseudoword Decoding	96.7 (13.7)	97.9 (16.0)	94.6 (14.2)	104.6 (9.3)	REF > TGA, TOF, SVF
Numerical Operations	96.8 (19.6)	95.7 (24.8)	93.4 (20.7)	110.7 (13.3)	REF > TGA, TOF, SVF
“Assembled” Composite	97.0 (16.6)	95.1 (20.9)	93.0 (18.7)	108.1 (10.9)	REF > TGA, TOF, SVF
Reading Comprehension	96.6 (18.3)	95.3 (20.7)	92.7 (19.4)	106.9 (13.0)	REF > TGA, TOF, SVF
Mathematics Reasoning	97.5 (17.6)	95.0 (23.2)	93.3 (20.6)	109.4 (11.7)	REF > TGA, TOF, SVF

Note: Data presented as n (SD); values reflect age-normed standard scores. Pairwise tests were adjusted for multiple comparisons.

Table 5

Hierarchical Multiple Regression Analyses Predicting Discrete/Associative and Assembled Academic Outcomes From CCHD Group and Visuospatial Organization and Integration Abilities

	Academic outcomes			
	Discrete/Associative		**Assembled**	
Predictor	**ΔR^2**	**β**	**ΔR^2**	**β**
Step 1	.292***		.263***	
Control variables[a]				
Step 2	.048***		.038***	
CCHD status[b]		.226***		.201***
Step 3	.051***		.050***	
DSS-ROCF Organization		1.075***		1.066***
Step 4	.004		.010**	
DSS-ROCF Style (Integration)		.064		.106**
Total Adjusted R^2	.386***		.353***	
n	452		452	

Note: CCHD = critical congenital heart disease.

[a] Control variables included age, SES, and DSS-ROCF Copy Basal Level

[b] Primary regression analyses compared CCHD vs. REF groups

$^{**}p < .01$.

$^{***}p < .001$.