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Neurocognitive functioning in adults with congenital heart disease

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Abstract

Objective: Adults with congenital heart disease (CHD) are at increased risk of psychological disorders and cognitive deficiencies due to structural/acquired neurological abnormalities and neurodevelopmental disorders as children. However, limited information is known about the neuropsychological functioning of adults with CHD. This study screened neuropsychological abilities and explored group differences related to cardiac disease severity and neurological risk factors in adults with CHD.

Design: Participants completed brief neuropsychological testing. Information about neurobehavioral and psychological symptoms, employment, education, and disability were also collected from the patient and a family member.

Results: Forty-eight participants with adult CHD completed neuropsychological testing. Visuospatial skills and working memory were worse than expected compared to the typical population. Frequency of neurological comorbidities (e.g., stroke, seizures) was higher in those with more severe heart disease (e.g., single ventricle or cyanotic disease), and executive functioning was weaker in those with neurological comorbidities. Those with more severe heart disease were more likely to be unemployed and to receive disability benefits, but educational attainment did not differ. Those who received disability performed worse on tasks of executive functioning.

Conclusions: Findings suggest concerns about neuropsychological functioning that need to be more comprehensively assessed in adults with CHD. Understanding the cognitive limitations of this aging population can help guide access to resources, transition of care, and medical care engagement, thus improving quality of care and quality of life.

KEYWORDS

adult congenital heart disease, adult transition, cognitive functioning, executive functioning, neurodevelopmental outcomes, neuropsychological outcomes

1 | INTRODUCTION

The increased risk of socioemotional, cognitive, and academic problems in children and adolescents with congenital heart disease (CHD) has been well-documented. Variability in outcomes is tremendous across the pediatric years, with subtle to severe concerns, and evolving challenges across the lifespan as demands increase. School age children may require grade retention, placement in the special education classroom, and they may be diagnosed with learning disabilities, attention-deficit/hyperactivity disorder, and executive functioning problems.[1–5] Compared with typical teens, they are unable to participate in competitive team sports and are not as physically active.[6,7] Social challenges and psychological problems continue into the teen years, and many need remedial supports to finish high school.[8] Quality of life is impacted across the lifespan for many of those with CHD.[9–11] In a large adult and geriatric survey study, dementia was reported to be the comorbidity of CHD with the highest magnitude, with a hazards ratio of 3.24.[12]

Variability in outcomes has been difficult to explain. Factors such as length of cardiopulmonary bypass,[13–15] circulatory arrest,[16,17] systemic venous oxygen saturations,[18] and type of procedure or surgery[19,20] do not consistently predict outcomes. Variables that are most consistently predictive of neurocognitive outcomes include length of hospital stay[19,21] and neurological events, such as seizures[8,16] or stroke.[22] Medical comorbidities also increase risk, such as a genetic condition with known neurodevelopmental problems.[23] Studies have also explored cardiac disease severity to help explain variable outcomes. Children and adolescents with single ventricle cardiac physiology[4,24] or congenital cyanotic disease tend to have worse outcomes.[25]

There are now more adults with CHD than children.[26] Considerable research in the adult CHD population shows an increased risk of mental health problems, especially internalizing symptoms.[27] For example, using a standardized clinical interview (Structured Clinical Interview for DSM Disorders), 50% of an adult sample (M = 39.1, SD = 11.3 years) met lifetime criteria for at least one mood or anxiety disorder, but 39% of these had never participated in mental health treatment.[28] The adult CHD population also presents with functional differences compared with the normative population. Adults (ages 30–43 years) with CHD had lower occupational status and yearly income, a higher percentage were living with their parents, and those with moderate to severe CHD experienced higher physical restrictions (e.g., play sports, perform physical labor), reduced physical strength, and endorsed feeling more at a disadvantage in life.[29] Those with severe CHD (i.e., single ventricle) compared with the general population had lower rates of employment, especially females.[30]

There is extremely limited information about the neuropsychological functioning (e.g., intelligence, language, nonverbal skills, memory, academic, sensory and motor, executive functioning) of adults with CHD. It is well documented that poor cardiovascular health (without CHD) can have significant consequences for neuropsychological functioning, ranging from mild cognitive changes to severe vascular dementia.[31] The typical profile for vascular cognitive impairment (without stroke) includes areas of executive functioning, such as processing speed, attention, learning efficiency, and memory retrieval.[32] One study estimated the intellectual and academic abilities of adults with CHD. Specifically, in a sample of adults (≥18 years) with severe CHD (e.g., single ventricle), intellectual and academic performance was around two standard deviations below the mean and significantly lower than those with less severe CHD.[33] Another study[34] with tetralogy of Fallot (27% cyanotic) assessed areas known to be more sensitive to neurological injury and abnormalities, such as executive functioning and memory. Descriptive findings explained that, compared to the normative population, "deficits" occurred for several tests of executive functioning involving working memory, sequencing and cognitive flexibility. A history of cyanosis was statistically associated with worse performance on some measures of executive functioning (e.g., cognitive flexibility and planning abilities).

Understanding the neuropsychological functioning of the aging patient with CHD will help facilitate access to community resources,[35] as well as inform ways to improve adult transition of care and problems related to medical adherence/engagement. The purpose of the current study was to assess the neuropsychological functioning of adults with CHD using a brief neurocognitive test battery. It was hypothesized that, compared to a normative sample, those with adult CHD would perform worse on measures of executive functioning, and worse on a rating scale that assessed psychological functioning. It was hypothesized that adults with single ventricle physiology would perform worse, compared with those who have two functional ventricles; and adults with cyanotic heart disease, compared with those who have acyanotic disease, would perform worse in executive and psychological functioning. Finally, it was also posited that adults with CHD who have a known neurological risk factor or comorbidity (e.g., stroke) compared with those with no known neurological history, would perform worse on tests of executive and psychological functioning.

2 | METHODS

2.1 | Participants

Forty-eight participants (17–56 years; males = 19) with CHD were recruited from an adult outpatient cardiology clinic at an academic medical center over a 2-year period. Patients were excluded from the study if they had acquired (not congenital) heart disease and/or if they were unable to understand consent or assent procedures. The ethnicity distribution of the sample was 77% Caucasian, 15% African American, and 8% other. See Table 1 for frequency of each cardiac diagnosis for the sample.

2.2 | Design and procedure

The current study was reviewed and approved by the Institutional Review Board. Informed consent was obtained from all participants. Consent from a legal guardian was required for one participant who was 17 years of age, and participant assent was also obtained for this individual. Due to length of time required for neurocognitive testing, a follow-up appointment was often scheduled as a separate visit from the clinic appointment. Participants completed a patient questionnaire that included information about education, employment, and disability services. Neuropsychological measures were administered following standardized procedures in a one-to-one setting. All testing was administered and scored by a psychometrician who was supervised by a licensed neuropsychologist. All scoring was checked for accuracy by a second psychometrician. The participant and a family member each completed separate standardized symptom rating scales during the testing session. Electronic medical records were reviewed by the study cardiologist (WMB) to clarify cardiac diagnosis. Medical records were also collected for genetic or neurological disorders and abnormal neuroimaging.

2.2.1 | Neuropsychological measures

A brief assessment of neuropsychological abilities included the following: The *Wechsler Adult Intelligence Scale, Third Edition* (WAIS-III)[36] Vocabulary, Block Design, and Digit Span Forward and Backward subtests; and *Delis-Kaplan Executive Functioning System* (D-KEFS)[37] Trails and Verbal Fluency subtests. The WASI-III Vocabulary subtest assesses oral vocabulary skills. The WASI-III Block Design subtest measures visuospatial constructional skills. The WASI-III Digit Span subtest

TABLE 1 Diagnoses by congenital heart disease group for the current sample

Cyanotic CHD	
Single Ventricle	
Hypoplastic left heart syndrome	2
Heterotaxy, single ventricle	2
Tricuspid atresia	3
Pulmonary atresia	1
Double inlet left ventricle (DILV)	2
DILV, pulmonary stenosis, Eisenmenger syndrome	1
Double outlet right ventricle (DORV)	1
Total SVC	**12**
Two Ventricle	
Pulmonary atresia	1
Tetralogy of Fallot	11
Atrial septal defect (ASD), Eisenmenger syndrome	1
Transposition of the great arteries (TGA)	5
TGA, Ventricular Septal Defect (VSD)	1
DORV	1
DORV, pulmonary stenosis	1
Total SVC	**21**
Acyanotic CHD	
Two Ventricle	
Coarctation of the aorta (COA)	3
COA, VSD	1
Aortic stenosis, bicuspid aortic valve	1
ASD	1
ASD, Patent ductus arteriosis	1
Ebstein's anomaly, triscuspid valve	1
Pulmonary stenosis	1
Dilated aortic root	1
Shone's syndrome, bicuspid aortic valve, COA	2
Ebsteins's anomaly	1
Congenital mitral stenosis	1
Infundibular pulmonary stenosis, VSD	1
Total SVC	**15**

assesses auditory working memory or online attention. The D-KEFS Trails assesses rapid sequencing and cognitive flexibility and D-KEFS Fluency measures rapid verbal retrieval and cognitive flexibility. Raw scores were transferred to standardized scaled scores (Scaled Score; mean = 10, SD = 3) using age-based norms available in the published test manual. Lower scores indicate worse performance compared with those of the same age from a normative sample.

The *Neurobehavioral Functioning Inventory* (NFI)[38] was completed by the patient and a family member to assess behaviors and symptoms that can be associated with neurological injury. The inventory is organized into six factor analytically derived scales: Depression, Somatic, Memory/Attention, Communication, Aggression, and Motor. Additional "critical items" were summed to assess high risk symptoms, including black out spells, seizures, threaten to hurt self, cannot be left home alone, miss/cannot attend work or school, and double/blurred vision. Raw scores were transferred to standardized T-scores and compared to a normative sample. Scores are interpreted as follows: $\leq$34 = Very Low, 35–43 = Low, 44–56 = Average, 57–65 = High, $\geq$66 = Very High. High symptoms are of clinical concern.

2.3 | Analytic plan

Shapiro-Wilk's test was performed in order to verify data distribution. With the normality of the data confirmed, descriptive statistics were carried out to characterize the sample. Comparison between the specified groups was analyzed in IBM SPSS Statistics, Version 20 (IBM Corporation, Armonk, NY, USA). Due to the large range in age, all analyses were run twice, once with age as a continuous covariate, and again without controlling for age. The most parsimonious model will be presented below, for which age was not a significant covariate, and thus removed from the results.

3 | RESULTS

3.1 | Descriptive and demographic findings for the sample

Severity of CHD for the sample was classified into three groups based on ventricle physiology (single- or two-ventricle circulation) and whether the CHD caused hypoxia (cyanosis or acyanosis). Groups included: "Single Ventricle Cyanotic" (SVC, n = 12), "Two Ventricle Acyanotic" (TVA, n = 15), or "Two Ventricle Cyanotic" (TVC, n = 21); all children born with a single ventricle have cyanotic disease (see Table 1). Regarding neurological comorbidities, 11 patients (23%) had a known neurological risk factor, which included abnormal neuroimaging, stroke, seizures, or a diagnosis of 22q11.2 deletion syndrome (i.e., DiGeorge syndrome), all of which can be associated with neuropsychological variability. Based on responses from a patient questionnaire, 30% of the sample was receiving disability, 13% had not and did not plan to go to college, 56% had planned or actually attended college, and 9% of the sample was unemployed. Refer to Table 2 for descriptive statistics of age, gender, and highest level of education separated by CHD group, Neuro Risk, and disability status. Analyses indicated that age, gender, and highest level of education did not differ based on CHD group or Neuro Risk. Although not significant, a somewhat larger proportion of adult CHD females than males are consistent with the literature.[39] A higher number of males compared to females reported receiving disability (see Table 2).

3.2 | Descriptive findings for NFI ratings

A series of correlations were conducted between the self-report and family member NFI ratings to assess the consistency in responses. Results indicate high consistency across most clinical scales for the self-report and family member NFI ratings for the TVA group; however, less consistency was observed in the SVC and TVC groups. All means were within the range of normal ($T \geq 57$ is clinically significant; Table 3). Regarding critical items, the frequency was low for both the NFI self-report and NFI family member ratings: blackout spells (n = 3 self/1 family), seizures (n = 1 self/1 family), threaten to hurt self (n = 1 self/0 family), cannot be left at home alone (n = 3 self/5 family), miss or cannot attend work/school (n = 9 self/9 family), double or blurred vision (n = 5 self/4 family). Findings highlight overall that both the patient and family member reported similar symptoms and that the current sample does not have significant elevations on this screening measure for psychological or neurological symptoms.

TABLE 2 Age, education, and gender based on CHD Group, Neuro Risk, and disability

	SVC		TVC		TVA		Analysis	
	N	Mean (SD)	N	Mean (SD)	N	Mean (SD)	*F* value	*P* value
Age	12	33.33 (12.77)	15	38.13 (8.53)	21	32.86 (10.51)	1.21	.31
Education	12	13.67 (2.87)	15	15.13 (2.61)	20	14.65 (2.39)	1.09	.34
Gender	12	M = 7, F = 5	15	M = 3, F = 12	21	M = 9, F = 12	$\chi^2 = 4.26$	.12
	Neuro Risk		No Neuro Risk				Analysis	
	N	Mean (SD)	N	Mean (SD)			*F* value	*P* value
Age	11	32.91 (9.6)	37	35.14 (10.9)			0.37	.55
Education	11	14.55 (2.8)	36	14.56 (2.57)			0.00	.99
Gender	11	M = 4, F = 7	37	M = 15, F = 22			$\chi^2 = 0.06$	.81
	Disability		No Disability				Analysis	
	N	Mean (SD)	N	Mean (SD)			*F* value	*P* value
Age	14	37.64 (11.64)	32	34.25 (9.46)			1.06	.31
Education	14	13.64 (2.06)	31	15.10 (2.48)			3.64	.06
Gender	14	M = 10, F = 4	32	M = 8, F = 24			$\chi^2 = 8.81$	.004*

Note: SVC, single ventricle cyanotic; TVA, two ventricle acyanotic; TVC, two ventricle cyanotic.
Education refers to self-reported highest number of years completed.
*$P \leq .01$.

3.3 | Descriptive findings for neuropsychological measures

One-sample *t* tests were then completed to assess whether the performance on neuropsychological measures for the current sample differed significantly from the mean score of the same–age normative population (Table 4). Results indicate that the current CHD sample performed worse on WAIS-III Block Design, WAIS-III Digit Span Forward, and WAIS-III Digit Span Backward. This indicates that, when compared with individuals of the same age in the general population, the current sample performed lower on measures of visuospatial construction and working memory (executive functioning). There were no differences between the CHD group means and the normative population for WAIS-III Vocabulary or the D-KEFS subtests. The current sample has oral vocabulary skills equivalent to that of the normative same-age population.

3.4 | CHD group differences for outcome measures

Given a priori hypotheses, a series of planned comparisons using Bonferroni correction for multiple comparisons, were conducted to explore the effect of CHD group on measures of neuropsychological functioning (WAIS-III subtests; D-KEFS subtests) and the NFI. The first planned comparison assessed whether patients with a cyanotic or acyanotic defect differed, regardless of whether they had a single or two ventricle diagnosis (TVC + SVC vs. TVA). There were no differences between these CHD groups on any neuropsychological measure (WAIS-III subtests; D-KEFS subtests), nor the NFI scales. The next planned comparison assessed whether group differences were present for CHD diagnosis based on number of ventricles (TVC vs. SVC). No differences were observed between these two groups for any neuropsychological measures (WAIS-III subtests; D-KEFS subtests) or the NFI rating scales.

3.5 | Neuro Risk, CHD group and outcome measures

Comorbid neurological and genetic conditions are a concern for those with CHD. Initial analyses showed that there were no differences between CHD groups for Neuro Risk, $\chi^2 = 3.35$, $P = .19$. In other words, for the entire sample, the frequency of having a neurological risk factor did not differ across CHD groups. Due to the small number of adults in the sample with neurological risk, additional analyses assessed CHD group difference for only those with neurological risk. Among the 11 participants with known neurological risk factors, there were group differences based on CHD disease severity (5 = SVC, 2 = TVA, 4 = TVC; $\chi^2 = 14.08$, $P < .01$). There were a higher number with Neuro Risk in the SVC and TVC groups compared to the TVA group.

A series of two-way analysis of variances (ANOVAs) were conducted to test for group differences in neuropsychological functioning and NFI scores between CHD groups and Neuro Risk groups. There was a significant main effect of CHD group for D-KEFS Trails Set Loss Errors, $F(2,37) = 6.00$, $P < .01$, partial $\eta^2 = .25$ (TVC = 78%, TVA = 90%, SVC = 100%), and D-KEFS Verbal Category Fluency, $F(2,37) = 3.92$, $P = .03$, partial $\eta^2 = .18$ (Scaled Score: TVC = 10.00, TVA = 11.20, SVC = 11.75). There was also a main effect of Neuro Risk on D-KEFS Verbal Category Switching Fluency, $F(1,37) = 6.96$, $P = .01$, partial $\eta^2 = .16$ (Scaled Score: No Neuro Risk = 10.84, Neuro Risk = 8.09). There was a significant interaction effect of CHD group and Neuro Risk on D-KEFS Verbal Fluency Category Switching Accuracy, $F(2,37) = 7.95$, $P = .001$, partial $\eta^2 = .30$ (Figure 1). This interaction highlights that performance is worse for those in the two ventricle CHD groups when

TABLE 3 Consistency between self-report and family member ratings on the neurobehavioral functioning inventory (NFI)

	Self		Family		Analysis	
	N	Mean (SD)	N	Mean (SD)	*r* value	*P* value
Single ventricle cyanotic						
Critical items	10	9.40 (2.12)	10	8.60 (2.84)	0.29	.42
Depression	10	41.40 (8.98)	10	38.90 (6.26)	0.74	.01**
Somatic	10	42.80 (6.49)	10	42.60 (6.33)	0.61	.06
Memory/Attention	10	42.60 (9.26)	10	38.40 (11.24)	0.64	.05*
Communication	10	46.40 (11.37)	10	41.70 (11.18)	0.74	.02*
Aggression	10	47.60 (8.75)	10	41.50 (5.28)	0.64	.05*
Motor	10	44.10 (7.40)	10	41.30 (7.90)	0.62	.06
Two ventricle cyanotic						
Critical items	16	7.88 (3.34)	16	7.44 (1.71)	0.26	.34
Depression	16	43.75 (8.24)	16	42.81 (9.33)	0.52	.04*
Somatic	16	44.25 (9.13)	16	40.56 (6.10)	0.46	.08
Memory/Attention	16	43.69 (7.55)	16	40.13 (9.28)	0.21	.44
Communication	16	49.75 (12.65)	16	45.75 (13.69)	0.59	.05*
Aggression	16	47.56 (9.95)	16	45.44 (9.41)	0.54	.03*
Motor	16	44.19 (8.86)	16	40.75 (8.59)	0.41	.11
Two ventricle acyanotic						
Critical Items	13	8.00 (2.24)	13	7.18 (1.47)	0.78	.001***
Depression	13	41.09 (6.96)	13	39.82 (6.98)	0.44	.14
Somatic	13	44.82 (5.86)	13	41.64 (5.97)	0.73	<.01**
Memory/Attention	13	41.45 (5.94)	13	36.82 (4.92)	0.82	.001***
Communication	13	42.91 (8.76)	13	38.27 (4.61)	0.64	.02*
Aggression	13	44.91 (5.87)	13	46.18 (7.31)	0.83	<.001***
Motor	13	42.18 (8.72)	13	38.09 (4.70)	0.68	<.01**

Note: Total raw score is used for Critical Items; T-Scores are used for all other clinical scales.
*≤ .05.
**≤ .01.
***≤ .001.

there are comorbid neurological risk factors. Finally, analyses indicated no group differences on the WAIS-III subtests or the NFI ratings.

3.6 | Disability status, Neuro Risk, CHD group, and outcome measures

The functional, day to day impact of CHD is also a concern. SVC patients were more likely to be unemployed or have limitations in their jobs compared to the TVC and TVA groups, χ^2 distribution = 17.55, $P = .03$ (Table 4). However, there were no differences between the groups for college attendance, with 8 of the 11 SVC patients attending college, 19 of the 21 TVC patients attending college, and 10 of the 14 TVA patients attending college. SVC patients were more likely to receive Disability services (χ^2 distribution = 7.89, $P = .02$), with 7/11 SVC patients on Disability, 5/21 TVC patients on Disability, and only 2/14 TVA patients on Disability. There was no difference between Neuro Risk and No Neuro Risk for receipt of Disability (Neuro Risk: 4 of 11 report receiving Disability; No Neuro Risk: 10 of 35 report receiving Disability).

Importantly, the patients that received Disability also performed significantly worse on executive functioning measures compared to patients who do not receive Disability. This included WAIS-III Digit Span Forward, $F(1,44) = 7.94$, $P < .01$ (Scaled Score: Disability = 7.28, No Disability = 9.46), WAIS-III Digit Span Backward, $F(1,43) = 6.49$, $P = .01$ (Scaled Score: Disability = 8.07, No Disability = 9.63), D-KEFS Trails Letter Sequencing, $F(1,44) = 5.94$, $P = .02$ (Scaled Score: Disability = 8.64, No Disability = 10.53), D-KEFS Trails Number Sequencing, $F(1,44) = 7.28$, $P = .01$ (Scaled Score: Disability = 8.36, No Disability = 10.63), and D-KEFS Trails Letter-Number Switching, $F(1,44) = 5.94$, $P = .02$ (Scaled Score: Disability = 7.86, No Disability = 10.25). There were no group differences in Disability for remaining subtests of the WAIS-III, D-KEFS, or NFI ratings.

4 | DISCUSSION

Individuals born with CHD are at high risk for structural/acquired neurological abnormalities and medical comorbidities that can affect the health of the brain, learning challenges, and social and emotional problems.[3,8] As adults, there are concerns about quality of life, psychological disorders, lower intellectual and academic functioning, lower employment and income, and higher rates of dementia.[11,12,33] Adults with CHD often have chronic, progressive cardiac disease and medical comorbidities can further complicate their life and their care. The first cohorts of children with CHD who have survived their palliative cardiac surgeries are now a large population of adults who need resources. These resources include assessment of risk (e.g., cognitive impairments and psychological disorders) and access to treatment and interventions to address risks that impact medical care engagement and quality of life.

The current findings from a brief neuropsychological assessment highlight concerns about executive dysfunction in adults with CHD,

TABLE 4 Performance on standardized neuropsychological measures compared with normative mean (Scaled Score = 10)

	N	Mean (SD)	*t* value	*P* value
WAIS-III Vocabulary	48	10.81 (2.92)	1.93	.06
WAIS-III Block Design	48	8.04 (2.16)	−6.27	<.01**
WAIS III Digit Span Total	48	9.35 (2.55)	−1.76	.08
WAIS-III Digit Span Forward	48	8.85 (2.85)	−2.79	<.01**
WAIS-III Digit Span Backward	48	9.11 (2.46)	−2.79	.02*
WAIS-III Digit Span Sequencing	47	9.83 (2.23)	−0.52	.6
D-KEFS Trails Visual Scanning	48	9.77 (3.22)	−0.49	.62
D-KEFS Trails Number Sequencing	48	9.90 (2.51)	−0.29	.61
D-KEFS Trails Letter Sequencing	48	9.79 (2.843)	−0.51	.61
D-KEFS Trails Switching	48	9.33 (3.29)	−1.40	.17
D-KEFS Trails Motor Speed	48	10.42 (2.07)	1.39	.17
D-KEFS Verbal Fluency Letter	48	9.44 (3.87)	1.01	.32
D-KEFS Verbal Fluency Category	48	10.81 (4.05)	1.39	.17
D-KEFS Verbal Fluency Switching	48	10.21 (3.60)	0.40	.69

Note: Wechsler Adult Intelligence Scale—3rd Edition (WAIS-III); D-KEFS, Delis-Kaplan Executive Functioning System. *$P \leq .05$, **$P < .01$.

especially when cardiac disease is more severe and there are comorbid neurological risk factors. Results support the study hypotheses and are consistent with outcome research in pediatric CHD and adult vascular disease without CHD.[4,32] Executive function includes cognitive processes related to attention, working memory (i.e., short-term memory), flexible thinking, planning, problem solving, and behavioral regulation (e.g., controlling impulsive behavior). These skills predict emerging independence at school and in the community throughout development.[40] Executive skills also predict adult independence in both healthy and clinical populations.[41,42] Problems with executive functioning likely contribute at least in part to functional differences in adults with CHD, including lower occupational status and not living independently from parents.[29] Indeed, findings from the current study highlight weaker executive functioning skills for those who received disability benefits. It is also likely that problems with executive skills contribute to barriers in ongoing cardiology care given the complexities of decision making and potential for emotional and cognitive overwhelm. This includes not wanting additional surgeries despite physician recommendations and poor understanding of cardiology needs.[43]

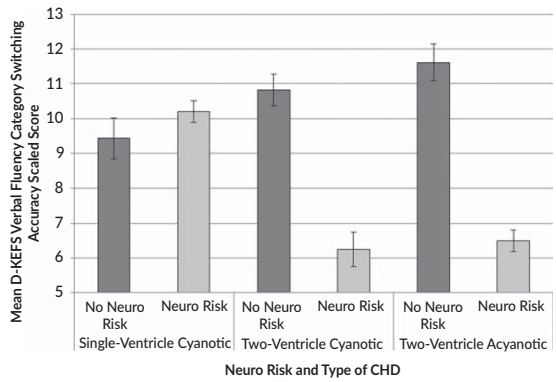

FIGURE 1 Interaction between CHD severity and neurological risk for cognitive flexibility on the Delis Kaplan executive function system (D-KEFS)

Addressing the psychological needs of the adult population with CHD is also essential, and has been highlighted in detail in other studies.[27,28] Although screening of select psychological symptoms (e.g., depression, aggression) in the current study was not clinically elevated, extensive research with more comprehensive assessment of psychological functioning indicates high incidence of depression and anxiety, with 40% seeking mental health treatment and 50% reporting interest in future psychological treatment.[44] Gathering psychological symptom reports from both the patient and a family members may be helpful in the clinic setting.

The limitations in the current study can guide future directions. First, the current sample may be biased toward those who experience more cognitive problems or have worse health/cardiac disease given that they were recruited from the clinic setting. Detailed information about those who declined to participate was not available; analyses to explore differences between those who decline or consent would help explain any sample biases. Additionally, it would be helpful to better understand the causes of disability in the current sample. Second, the current neuropsychological test battery was brief. Future studies will want to explore neuropsychological functioning more comprehensively. A thorough assessment of executive functioning, memory, and visuospatial processing will be especially important given that these areas tend to be weak in pediatric CHD and vascular diseases. Third, a broad age range was recruited for the current sample; in the future, a larger

sample and a more focused age group may help improve variability and thus the ability to statistically detect group differences. Limited information about medications, comorbid conditions, and neuroimaging was available through record review and patient report. A more thorough, prospective collection of potential medical covariates will be important. This might include the patient's history of neurological events (e.g., seizures, stroke), current medications that may impact cognitive functioning, current oxygen saturation levels, and number/type of cardiac surgeries. Finally, longitudinal assessment of the neuropsychological abilities and neurological health (using MRI) of the aging patient with CHD will be essential. The implications of chronic hypoxia and progressive cardiac disease on the health of the brain in an adult with CHD is unknown to date.

The findings from this study have clinical implications. Cardiology providers will need to consider the impact of cognitive weaknesses and impairments on patient decision making, engagement in care, adherence, and quality of life. Those with severe or progressive CHD and neurological/genetic risk factors are at higher risk and should be monitored closely for referrals to specialists. For example, young adults who are struggling with independence or who present with cognitive/psychological concerns should be referred to a psychologist or counselor in the community for intervention and access to resources. If the aging adult with CHD presents with cognitive or memory concerns, the patient and family should be referred to an adult neuropsychologist for comprehensive assessment and clinical management (e.g., neuroimaging follow-up, cognitive remediation, community rehabilitation, and psychological/family counseling referrals). With executive functioning impairments, patients might be inattentive, slow to process and learn information (needing repetition), disorganized, forgetful, avoidant of time consuming or complex tasks/decisions, inconsistent, impulsive, demonstrate poor planning/problem solving, and lack self-awareness about their difficulties. These patients may need more reminders, help with organizing/scheduling, as well as more extensive and repeated education during their medical visits. Given the complexities of the aging patient with CHD, building and maintaining relationships with community providers and specialists is essential to our ability to maximize quality of medical care and quality of life for patients and families.

5 | CONCLUSIONS

Little is known about the neuropsychological functioning of the adult patient with CHD, despite their risk for problems that can impact quality of life and independence. Findings from the current study revealed visuospatial skills and working memory that were lower than expected compared to the typical population, and executive functioning was weaker in those with neurological comorbidities compared to those without. Those with more severe heart disease were more likely to be unemployed and to receive disability benefits, but educational attainment did not differ. Those who received disability performed worse on measures of executive functioning. High risk adults with CHD, such as those with severe cardiac disease and those with neurological risk factors, need to be identified as at high risk for neuropsychological deficiencies and be referred appropriately for mental health and neuropsychological services. Future directions should include more comprehensive, longitudinal assessment of the neuropsychological functioning in the aging adult with CHD.

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CONFLICT OF INTEREST

The authors of this manuscript have no conflict of interest to disclose.

AUTHOR CONTRIBUTIONS

DI assisted with concept/design, data analyses/interpretation, and drafting of article; KEO assisted with concept/design, data analyses/interpretation, and drafting of the article; RM assisted with concept/design and data collection; WB assisted with concept/design and drafting of article; AYS assisted with concept/design, data analyses/interpretation, drafting of article.

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