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ORIGINAL ARTICLE

WILEY Congenital Heart Disease

Identifying self-reported neurocognitive deficits in the adult with congenital heart disease using a simple screening tool

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Abstract

Objective: Children with congenital heart disease (CHD) and adults with acquired heart disease are at an increased risk of neurocognitive impairment. The objective of this study was to determine the prevalence of self-reported neurocognitive impairment and its risk factors in the adult congenital heart disease (ACHD) population.

Design: The Wisconsin Adult Congenital Heart Disease Program recently began screening ACHD patients to identify those with significant self-perceived neurocognitive impairments. Screening consists of using a validated neuro-oncology screening instrument that has been modified for the ACHD population. Patients who answer this survey in a predetermined fashion consistent with significant self-perceived neurocognitive deficits are referred for a formal neurocognitive evaluation. Demographic and clinical information are obtained by chart review.

Results: Three hundred ten patients (49% males) completed the screening process. The average age was 30 years (range: 17–69 years). For the cohort, 57 (18%) patients had no prior cardiac surgeries, 85 (28%) one surgery, 77 (25%) two, and 91 (29%) at least three surgeries. Of those screened, 106 (34%) met criteria for a formal neurocognitive evaluation. Patients who were referred had undergone a greater number of prior cardiac surgeries (2.2 vs 1.7, $P = .008$) and were more likely to have severe complexity CHD ($P = .006$). Of those patients who were referred, the worst perceived functioning was in math and attention.

Conclusion: There is a high prevalence of ACHD patients with significant self-perceived neurocognitive deficits. Simple screening questionnaires may help identify those patients at high risk and allow for timely and appropriate referral for formal neurocognitive evaluation, diagnosis, and therapy.

KEYWORDS
congenital heart disease, neurocognitive deficit

1 | INTRODUCTION

Cognitive function is defined as an individual's cerebral activity by which one perceives, comprehends, and understands the world around them. It is traditionally divided further into distinct cognitive domains, such as: intelligence, language, learning, memory, motor function, attention, and executive function.[1] Neurocognitive function in children with congenital heart disease (CHD) has been well studied in multiple studies over the last 20 years. These studies have identified a distinct pattern in children with congenital heart disease characterized by mild cognitive delay as measured by intelligence quotient, deficits in language (in particular pragmatic

language), poor psychomotor skills (gross, fine, and visual-motor), inattention with an increased risk for attention deficit hyperactivity disorder, impaired executive function, and impaired social interaction.[1–3] Adults with acquired heart disease, such as heart failure, are also likely to have cognitive impairments in multiple domains. These cognitive deficits are felt to be due to a decrease in cerebral blood perfusion leading to a loss of gray mater. These deficits can place an individual at risk of an inability to care for oneself, manage one's dietary restrictions, and one's medication regimen. This may lead to an increased risk of hospitalization and an increase in mortality.[4–6]

In the adult survivor with congenital heart disease, there remain multiple unanswered questions in regard to neurocognitive function, including the impact of neurocognitive deficits on patient-centered outcomes such as quality of life. Finally, the best method for detecting neurocognitive dysfunction in the ACHD patient to allow early intervention is unknown. In efforts to begin answering some of these questions, at our center, we sought to identify the prevalence and risk factors for self-reported neurocognitive deficits in our ACHD patients using a simple, easily administered screening tool.

2 | METHODS

2.1 | Study design and population

Between February 2016 and December 2016, the Wisconsin Adult Congenital Heart disease program (WAtCH) conducted a quality improvement project screening patients 18 years of age or greater with CHD for self-reported neurocognitive deficits. Patients who had a known genetic syndrome, neurocognitive deficit, or patients that did not have CHD were excluded. Because this project was approved by the Children's Hospital of Wisconsin's (CHW) Institutional Review Board (IRB) as a quality improvement project, the need to obtain informed consent was waived. This study protocol conforms to the ethical guidelines of the 1975 Declaration of Helsinki as reflected in a priori approval by the CHW IRB.

2.2 | Screening

All patients were screened for self-reported neurocognitive deficits using a modified version of a previously validated neuro-oncology neurocognitive screening questionnaire (FACT Brain).[7,8] The questionnaire was administered in paper form and could be completed in less than 10 minutes (Figure 1). This questionnaire consisted of 10 questions answered on a 5-point Likert scale that measures a subject's self-perceived competence in eight neurocognitive domains (attention, memory, language, executive functioning, independence in activities of daily living, math, processing speed, and reading). The subject could answer each question on a scale of 0–4. An answer of 0 corresponded to a profound deficiency, while an answer of 4 corresponded to no disability in the given domain. Based on patient responses, the study cohort was further divided into those patients who self-reported significant neurocognitive deficits and those patients who self-reported no or only minor neurocognitive deficits. After discussion with our collaborating neuropsychologist, it was determined that patients with significant self-reported neurocognitive deficits would be denoted by answering a "2" or lower on any two questions, or by answering the only question related to memory as "2" or lower. These patients were then referred for a formal neurocognitive evaluation. In addition to survey administration, a detailed chart review was performed allowing for analysis of risk factors. Data collected included age, sex, primary CHD diagnosis,

Neurocognitive Screening Survey

Name: **Age:** **Date:**

By circling one (1) number per line, please indicate how true each statement has been for you during the past 7 days.

	Not at All	A Little Bit	Some-what	Quite a bit	Very much
1. I am able to concentrate and/or sustain my attention	0	1	2	3	4
2. I can remember new things	0	1	2	3	4
3. I am independent in routine responsibilities (e.g., finances, driving, cooking, taking meds)	0	1	2	3	4
4. I can find the right word(s) to say what I mean	0	1	2	3	4
5. I am able to express my thoughts	0	1	2	3	4
6. I am good at math	0	1	2	3	4
7. I am able to organize/plan/multitask	0	1	2	3	4
8. I am able to think quickly	0	1	2	3	4
9. I have good reading comprehension skills	0	1	2	3	4
10. I am good at decision making and problem solving	0	1	2	3	4

FIGURE 1 The modified version of the FACT Brain used as the neurocognitive screening tool

age of complete repair in patients, number of prior heart surgeries, CHD complexity, and medications.

2.3 | Statistical Analysis

The outcome of neurocognitive deficits was analyzed using chi-squared tests for categorical variables and *t* tests for continuous variables. Descriptive statistics were also performed. A *P* value of ≤ .05 was considered significant.

3 | RESULTS

3.1 | Baseline characteristics

Table 1 shows baseline demographic and clinical characteristics for the entire cohort, those who self-reported significant neurocognitive deficits, and those with minor or no deficits. A total of 337 patients (50% females) were screened, mean age 30 years (SD: 10.3 years). Of the 337 patients, 116 (34%) patients self-reported significant neurocognitive deficits (Figure 2). Compared to patients who reported minor or no deficits, patients who reported significant deficits had similar baseline demographics: based on age (31 ± 10.8 years vs 30 ± 10 years, *P* = .44) and gender (53% females vs 51% females, *P* = .51). Patients who had significant self-reported neurocognitive deficits however were more likely to have severe complexity CHD (*P* = .003), were more likely to have had undergone a greater number of prior surgeries (2.1 ± 1.7 vs 1.7 ± 1.3, *P* = .014), and had a strong trend toward having a longer duration of cyanosis (6.2 years vs 3.2 years, *P* = .06).

TABLE 1 Baseline characteristics

Demographic	Total cohort (N = 337)	Referred (N = 116)	Not referred (N = 221)
Age (years)	30 ± 10.3	31 ± 10.8	30 ± 10.0
Female (%)	50%	47%	51%
Born with cyanotic CHD (%)	43%	48%	40%
Duration of cyanosis (mean years)	4.4	6.2	3.3
Number of prior surgeries (mean)	1.88 ± 1.5	2.1 ± 1.7	1.7 ± 1.3
Severe complexity CHD (%)	34%	44%	28%
Medications:			
Beta-blockers (%)	29%	43%	22%
ACE-I/ARBS (%)	28%	40%	22%
Digoxin (%)	9%	17%	5%
Spironolactone (%)	8%	17%	3%
Aspirin (%)	33%	47%	27%
Antiarrhythmics (%)	6%	17%	2%
Diuretics (%)	8%	17%	3%

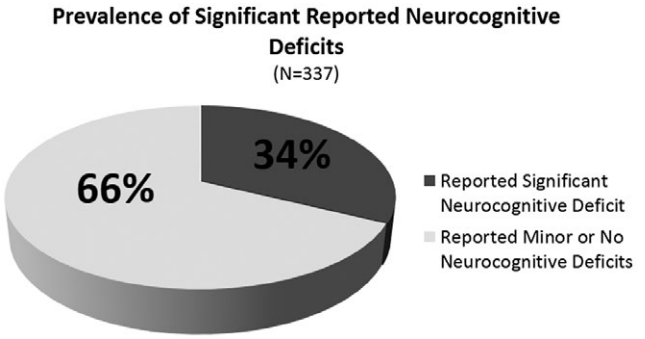

FIGURE 2 Prevalence of significant reported neurocognitive deficits. Pie chart showing the prevalence of patients who self-reported significant vs minor or no neurocognitive deficits

3.2 | Neurocognitive function

Figure 3 shows the mean scores for both cohorts in each of the eight neurocognitive domains. Those who self-reported significant deficits had worst perceived cognitive function in all eight of the cognitive domains compared to those who self-reported minor or no perceived deficits. This difference was most striking in the domains of math, attention, and memory. Their best perceived cognitive function was in executive functioning and independence in activities of daily living, but remained significantly worse compared to the cohort who reported no or only minor deficits.

Of the 116 patients who self-reported significant neurocognitive dysfunction and were referred for formal neurocognitive testing, only 20 patients thus far have completed formal neurocognitive testing. Of the remaining 96 patients, 46 patients declined appointments after being contacted directly by the neurocognitive office for personal choice. An additional 40 patients scheduled their neuropsychology testing appointment but then no-showed. The remaining 10 patients have moved out of state, were unable to schedule due to insurance issues, or unable to return due to travel distance. Of the 20 patients (18%) who have completed their formal evaluation, 18 of these patients (90%) were found to have significant neurocognitive deficits that were felt to necessitate referral to appropriate therapy and counselors. The remaining two had minor deficits and were given counseling but were felt not to warrant further intervention or therapies. Interestingly, patients who have completed formal neurocognitive testing were more likely to have more severe complexity CHD compared to those that did not complete their neurocognitive evaluation (*P* = .037).

4 | DISCUSSION

To date, there only exist eight studies in the literature that specifically evaluate neurocognitive dysfunction in the adult with congenital heart disease. The prior literature has been limited by small sample sizes (less than 60 patients) and the use of a variety of different modalities to measure cognitive dysfunction. In our study, we

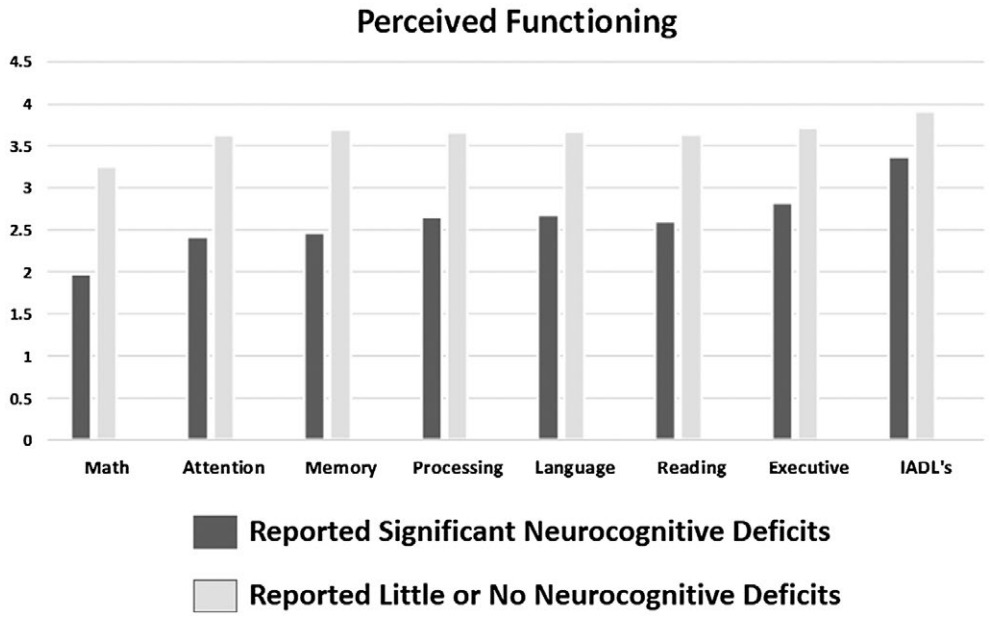

FIGURE 3 Perceived neurocognitive functioning. This chart shows how both patients that reported significant neurocognitive deficits (dark grey) and those that did not report significant neurocognitive deficits (light grey) perceived their function in each of the eight neurocognitive domains listed on the x-axis. The y-axis represents the average Likert score for each domain, remembering that a lower score indicates a worse perceived

used a modified version of a well-validated neurocognitive screening questionnaire to screen a large heterogeneous ACHD population. This questionnaire was easy to administer and took less than 10 minutes. Overall, our study is the largest published to date and demonstrates that over a 1/3 (34%) of ACHD patients screened in a busy ACHD program self-report significant neurocognitive dysfunction. This dysfunction was most perceived in the cognitive domains of math, attention, and memory, but these high-risk patients reported worse dysfunction in all domains compared to patients who had only minor or no reported deficits. Risk factors associated with self-perceived significant neurocognitive impairment included: severe complexity of CHD and number of prior cardiac surgeries. Unfortunately, we had a low rate of formal neurocognitive evaluation (18%); however, for those that did complete their evaluation, 90% of them were found to have a significant neurocognitive deficit and were given recommendations and referred to therapy to address the deficit. These recommendations often centered upon recommending visiting job training programs, vocational rehab centers, and academic disability resource centers where patients can seek academic adjustments (scribes, tutoring, etc). Interestingly, those that did follow through and complete their formal evaluation were more likely to have severe complexity CHD which may indicate a higher awareness of their neurocognitive deficits, having a deficit that significantly impacts their quality of life, or a stronger motivation to find a strategy to overcome these deficits.

Our study adds to the growing literature demonstrating a significant presence of neurocognitive impairments in the ACHD population. Ilardi et al found that by using a few brief neuropsychological tests, 48 ACHD patients had overall worse visuospatial skill and working memory compared to age-based norms, and that those with severe congenital heart disease were more likely to be unemployed and to receive disability.[9] Daliento et al found that by using a well-validated neuropsychological battery, 54 ACHD patients with a history of repaired tetralogy of Fallot showed an overall increased prevalence of deficits in executive function, problem solving, and planning. This study cohort was also noted to have a lower than normal academic level and more jobs inadequate for educational level.[10] Klouda et al using a computerized battery of standardized neurocognitive tests (CNS-Vital Signs) found that out of 48 ACHD patients, those with severe CHD had worse processing speed, attention, and executive function. Like our study, Klouda also found that the number of prior surgeries had a strong relationship to neurocognitive deficits, specifically to worse executive function.[11]

Despite the increasing awareness of neurocognitive deficits in the ACHD population, there is little research regarding the impact of these deficits on our patients' lifestyle and quality of life. Apers et al demonstrated that educational attainment, employments status, and marital status were predictors of quality of life in ACHD patients,[12] and other prior studies have noted that ACHD patients tend to have worse educational attainment, more unemployment, and be less likely to be in a significant relationship or married.[13-16] Future studies will be needed to delineate when the optimal age for screening the ACHD population for neurocognitive deficits may be. However, early screening for and identification of neurocognitive deficits in the ACHD population may help tailor patient-specific interventions that may improve employment status, educational achievement, and possibly the ability to maintain relationships to ultimately improve our patients' quality of life.

Unfortunately, there currently is no validated neurocognitive screening survey specifically for the ACHD population. Previous studies, however, have demonstrated that surveys screening for self-perceived neurocognitive deficits reliably predict neuropsychological impairment in other disease populations, such as multiple sclerosis.[17] In this study, we used a modified version of a well-validated neurocognitive screening instrument (FACT Brain) to screen our patient population. While the FACT Brain has been well validated and used frequently in the neuro-oncology patient population, this specific version of the screening instrument has not been used in the ACHD population. As a result, it is possible that we overestimated the presence of neurocognitive dysfunction in our cohort (low specificity). For this reason, all patients who self-reported significant dysfunction were referred for formal neurocognitive testing to confirm the presence of neurocognitive dysfunction and as a result help validate our findings. Thus far, only 20 patients have undergone formal testing, but of these, 90% have been found to have significant deficits necessitating referral for further therapies and interventions. While this is a limitation, it is not overly surprising that patients with the more complex congenital heart disease and who would likely benefit the most from possible therapies and interventions are the ones that followed through with a full evaluation. The patients that were more likely to not follow through with a formal evaluation were those with simple congenital heart disease. This may reflect that the screening tool has a low specificity, or perhaps these patients have a better ability to compensate for their neurocognitive deficits and did not feel they needed a formal evaluation. Some may also consider this an example of poor executive function in of itself and a positive screen. Larger studies and more outcomes of formal neurocognitive evaluations are needed to better delineate whether routine screening of simple congenital heart disease is needed.

5 | LIMITATIONS

Another limitation to our study was that this was a single center study. In addition, not all domains in the screening tool were represented equally with some neurocognitive domains evaluated with only one question. All questions were worded in the positive but there was no validity indicator to raise suspicion of individuals who tend to say either yay or nay. We did not assess relationships between neurocognitive deficits and quality of life predictors, such as: educational attainment, employment status, and relationship status. Large, multicenter studies are needed to determine the associations between neurocognitive deficits and quality of life.

6 | CONCLUSION

Despite these limitations, we feel that our simple neurocognitive screening tool is useful for assessing ACHD patients for self-reported neurocognitive deficits. While further studies are needed to fully validate this tool in the ACHD population, our data again support that neurocognitive deficits in the ACHD patient are common. Preliminary data indicate that these deficits clearly play a role in our patient's ability to achieve a high quality of life. For this reason, early screening for and identification of neurocognitive deficits in the ACHD population may help tailor patient-specific interventions that may improve our patients' quality of life.

CONFLICT OF INTEREST

There are no conflicts of interest to disclose.

AUTHOR CONTRIBUTIONS

Ashley Brunmeier MD: Concept and design, Data collection, Drafting article, Critical revision of article, and Approval of article.

Michael P Reis BS: Data collection and Drafting article.

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Salil Ginde MD: Drafting article, Critical revision of article, and Approval of article.

Peter J. Bartz MD: Critical revision of article and Approval of article.

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