

Nearly Half of Chiari Patients Show Signs of Cerebellar Cognitive Dysfunction

Cognitive problems such as brain fog, difficulty concentrating, memory problems, and trouble finding words are commonly reported by people with Chiari. A new study approached these problems from a different angle by using a test specifically designed to detect cognitive dysfunction related to the cerebellum.

The researchers administered the Cerebellar Cognitive Affective Syndrome (CCAS) Scale to 73 symptomatic Chiari patients ranging in age from 8 to 60. The group included 56 adults and 17 children, and 82% were female. Forty-three of the patients eventually underwent decompression surgery, while 30 were managed without surgery. CCAS, sometimes called Schmahmann syndrome, refers to a pattern of cognitive and emotional problems that can occur when the cerebellum is affected. While the cerebellum was once thought to primarily control movement and balance, research over the past several decades has shown that it is also connected to brain networks involved in attention, language, memory, planning, and emotional regulation. The CCAS Scale includes 10 short tests covering areas such as verbal fluency, attention, working memory, verbal recall, reasoning, impulse control, visuospatial ability, and affect. Failing one test is classified as "possible" CCAS, two as "probable," and three or more as "definite."

In this Chiari group, 44% of the patients met the criteria for definite CCAS, meaning they failed at least three of the ten tests. Another 15% met the criteria for probable CCAS and about 29% for possible CCAS. Only about 12% of the patients had no failed tests. The most common problem was with Digit Span Forward, a test of attention and immediate verbal memory in which a person repeats increasingly long strings of numbers. Nearly half of the entire group failed this test.

CCAS Test	Percent Who Failed
Digit Span Forward	46.6%
Phonemic Fluency	28.8%
Digit Span Backward	28.8%
Similarities	28.8%
Verbal Recall	27.4%
Category Switching	26.0%
Cube Copy	26.0%
Go/No-Go	19.2%
Semantic Fluency	16.4%

The pattern is interesting because the greatest difficulty was seen in attention, short-term memory, and several executive-language tasks, while simpler semantic and visuospatial abilities were relatively less affected. The researchers also compared CCAS scores with standard neuropsychological testing in a subset of 28 adults. Overall CCAS performance was significantly related to broader measures of intelligence, academic skills, and executive function, lending some support to the idea that the shorter CCAS test is measuring meaningful cognitive abilities. Several individual CCAS subtests also correlated strongly with comparable tests from standard neuropsychological batteries.

Fifty-three percent of the children met criteria for definite CCAS compared with 41% of the adults, but the CCAS Scale is not adjusted for age. The authors acknowledge that the poorer performance among children is likely influenced by normal developmental differences in executive function and language ability.

Interestingly, overall CCAS scores did not correlate with overall surgical outcome scores, and patients classified as possible, probable, or definite CCAS did not differ significantly in their outcomes.

The study adds to growing evidence that cognitive symptoms in Chiari likely involve more than the indirect effects of pain, poor sleep, or emotional distress and may, at least in some patients, reflect dysfunction in cerebellar-cerebral brain networks.

Source: Amaral-Nieves N, Shao B, Tiemeyer ME, et al. The CCAS-Scale in Chiari Malformation Type I: A Cerebellar-Focused Cognitive Assessment with Clinical Utility. *The Cerebellum*. 2026;25:127. doi:10.1007/s12311-026-02072-5

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