Journal of the International Neuropsychological Society (2025), 1–11
doi:10.1017/S1355617725101033

Research Article

Validation of the cerebellar cognitive affective syndrome (CCAS) scale in CCAS patients and cerebellar controls

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Abstract

Objective: The cerebellar cognitive affective syndrome (CCAS) scale has been developed to screen for possible cognitive and affective impairments in cerebellar patients, but previous studies stressed concerns regarding insufficient specificity of the scale. Also, direct comparisons of CCAS scale performance between cerebellar patients with and without CCAS are currently lacking. The aim of this study was to evaluate the validity of the CCAS scale in cerebellar patients. **Method:** In this study, cerebellar patients with CCAS (n = 49), without CCAS (n = 30), and healthy controls (n = 32) were included. The Dutch/Flemish version of the CCAS scale was evaluated in terms of validity and reliability using an extensive neuropsychological assessment as the gold standard for CCAS. Correlations were examined between the CCAS scale and possible confounding factors. Additionally, a correction for dysarthria was applied to timed neuropsychological tests to explore the influence of dysarthria on test outcomes. **Results:** Cerebellar patients with CCAS performed significantly worse on the CCAS scale compared to cerebellar controls. Sensitivity was acceptable, but specificity was insufficient due to high false-positive rates. Correlations were found between outcomes of the scale and both education and age. Although dysarthria did not affect the validity of the CCAS scale, it may influence timed neuropsychological test outcomes. **Conclusions:** Evaluation of the CCAS scale revealed insufficient specificity. Our findings call for age- and education-dependent reference values, which may improve the validity and usability of the scale. Dysarthria might be a confounding factor in timed test items and should be considered to prevent misclassification.

Keywords: Cerebellar cognitive affective syndrome; cognition; affect; validation; cerebellum; ataxia

(Received 17 December 2024; final revision 13 March 2025; accepted 17 April 2025)

Statement of Research Significance

Research Question(s) or Topic(s):

- Cognitive and affective symptoms in patients with cerebellar disorders are referred to as the cerebellar cognitive affective syndrome (CCAS).
- To evaluate cognitive deficits as part of CCAS, a brief screener – the CCAS scale – was developed.
- Considering indications of suboptimal validity, this study aimed to evaluate the validity of the CCAS scale using a gold standard neuropsychological examination and a control group of cerebellar patients.

Main Findings:

- Sensitivity of the CCAS scale was acceptable, but specificity was insufficient due to high false-positive rates.
- Correlations were found between outcomes of the scale and both education and age.

Study Contributions:

- Our findings call for age- and education-dependent reference values, which may improve the validity and usability of the scale.

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Cite this article: Reumers S.F.I, Maas R.P.PW, van den Brandt V.J.M, Kocken L.A.H, Wiegand I.M., de Leeuw F.-E., Schutter D.J.LG, van de Warrenburg B.P.C, & Kessels R.P.C. Validation of the cerebellar cognitive affective syndrome (CCAS) scale in CCAS patients and cerebellar controls. *Journal of the International Neuropsychological Society*, 1–11, https://doi.org/10.1017/S1355617725101033

Introduction

Patients with cerebellar disorders typically have motor- and vestibular-related dysfunctions like ataxia, but also commonly have cognitive and affective symptoms (Hernández-Torres et al., 2021; Mak et al., 2016; Reumers et al., 2024, 2025). This is seen as the third cornerstone in clinical ataxiology and is referred to as the Cerebellar Cognitive Affective Syndrome (CCAS) (Manto & Mariën, 2015; Schmahmann & Sherman, 1998). CCAS has been reported in a wide variety of cerebellar disorders, including those with structural lesions and degenerative ataxias (Hadjivassiliou et al., 2017; Ramirez-Zamora et al., 2015). The syndrome is characterized by impaired executive function, disturbed spatial cognition, personality changes, and language deficits (Schmahmann & Sherman, 1998). In recent years, the neurocognitive and affective profile has been described in more detail, including deficits in information processing speed, response inhibition, verbal fluency and memory, behavioral problems, and emotional disturbances (Ahmadian et al., 2019; Hoche et al., 2018; Wolf et al., 2009). Evaluating impairments as part of CCAS usually requires an extensive neuropsychological assessment, as standard screening instruments such as the Mini-Mental State Examination (MMSE) or the Montreal Cognitive Assessment (MoCA) have a more general focus and are not specific to cerebellar deficits. Since patients usually perform in the clinically unimpaired range on these screeners, the lack of sensitivity makes them not suitable to differentiate between CCAS patients and controls (Alan et al., 2024). However, extensive neuropsychological assessment is time-consuming and not always feasible in clinical practice. Therefore, a brief and easy-to-administer bedside tool – the CCAS scale – was developed to examine cognitive and affective functioning in cerebellar patients (Hoche et al., 2018).

The original CCAS scale has been developed (n = 77) and validated (n = 39) in a US cohort and included patients with isolated cerebellar or complex cerebrocerebellar disorders (Hoche et al., 2018). Outcomes were categorized into "possible", "probable", and "definite" CCAS, with a sensitivity of 46–95% and specificity of 78–100% for the different outcomes. The scale was recently evaluated in a larger US sample (n = 309), describing a sensitivity of 46–83% and specificity of 46–95% (Selvadurai et al., 2024). The CCAS scale has been translated into several other languages, and diagnostic properties have been evaluated in cohorts comprising different etiologies (de Oliveira Scott et al., 2023; Guo et al., 2024; Maas et al., 2021; Rodríguez-Labrada et al., 2021; Szabó-Műhelyi et al., 2024; Thieme et al., 2020, 2022; Van Overwalle et al., 2019). Results of these studies indicate suboptimal discriminative ability between patients and controls due to high false-positive rates, and attempts have been made to obtain more optimal threshold values. The issue of high false-positive rates may be caused by the fact that the scale does not take the effects of age and educational level into account (Thieme et al., 2021). Furthermore, the role of dysarthria in the assessment of CCAS-related deficits has recently been stressed (Corben et al., 2024). Dysarthria may overestimate cognitive verbal fluency deficits and thereby contribute to false-positive outcomes. However, several studies indicate the presence of language deficits even after dysarthria was taken into account, suggesting that poor test performance may be partially, but not fully explained by dysarthria (Cocozza et al., 2018; Stoodley & Schmahmann, 2009).

Despite indications of suboptimal validity, the scale was already used in multiple studies to describe CCAS in specific etiologies or assess its prevalence (Abderrakib et al., 2022; Bolzan et al., 2024; Destrebecq et al., 2023; Destrebecq & Naeije, 2023; Dujardin et al., 2024; Naeije et al., 2020; Selvadurai et al., 2024). Using the CCAS scale as a reliable screen presents a challenge as it has not yet been validated against extensive neuropsychological assessment, which is considered the gold standard for identifying cognitive dysfunction (Lezak et al., 2012). However, there is no consensus on the exact set of neuropsychological tests to establish or refute a CCAS diagnosis. Studies evaluating the scale thus far involved cerebellar patients without further specifying whether these individuals actually fulfilled the criteria of CCAS. It is possible that not all cerebellar patients have CCAS, or that a substantial proportion of patients have a mild cognitive impairment, as recently indicated in a large multicenter study (Liu et al., 2024). Demonstrating that the CCAS scale discriminates between cerebellar patients and a healthy control group is insufficient to determine the validity, as in clinical practice it will be used to distinguish between cerebellar patients with CCAS and those without. Therefore, a comparison with a gold standard to distinguish patients with and without actual CCAS is needed to establish the scale's diagnostic accuracy and clinical value.

It is evident that a valid CCAS screen is essential to accurately identify cognitive and affective deficits in persons with cerebellar disorders, both in neuropsychological research and clinical practice (Chirino-Pérez et al., 2021; Gok-Dursun et al., 2021; Kotkowski et al., 2021). Validation and possible improvement of the scale, considering the aforementioned restraints, is hence important. In this study, we will examine two research questions: (1) what is the validity of the CCAS scale when compared to a control group of cerebellar patients, using a gold standard neuropsychological examination? And (2) what is the influence of dysarthria on neuropsychological test outcomes and CCAS scale validity?

Method

Participants and procedure

Data were collected at the Neurology department of Radboud University Medical Center and the Donders Centre for Cognition (DCC) in the Netherlands. Three groups of participants were included: (1) cerebellar patients with CCAS, as determined by an extensive neuropsychological examination, (2) cerebellar patients without CCAS, and (3) healthy controls (HC). Cerebellar patients included all types of degenerative cerebellar ataxias and cerebellar strokes. Minimum required sample sizes were 40, 30, and 30, respectively, as determined by a power calculation for receiver operating characteristic (ROC) analysis (power > 0.9, AUC > 0.8) with an alpha level of significance of 0.05 (two-tailed). Cerebellar patients were part of a randomized controlled trial with CCAS patients in the Netherlands (Dutch Trial Register: NL9121) or visited the outpatient clinic of the neurology or rehabilitation departments of our center. Since we did not know in advance whether the cerebellar patients would have CCAS or not, we unintentionally included more CCAS patients than the predetermined sample size. The HC group was recruited through a pool of healthy volunteers at the DCC who participated in a larger neuropsychological test battery. All eligible participants were 18 years or older and fluent in Dutch. Exclusion criteria were any (comorbid) neurological or psychiatric disorders (self-report). This study was approved by the medical ethics committee (CMO Arnhem-Nijmegen, 2021-8296), as well as the trial (CMO Arnhem-Nijmegen, NL73572.091.20) and the study at the DCC (ECSW2017-2306-520). This study was preregistered (AsPredicted#147624), and all participants provided written informed consent before inclusion, in accordance with the declaration of Helsinki.

https://doi.org/10.1017/S1355617725101033 Published online by Cambridge University Press

Table 1. Test items of the test battery and CCAS scale, with corresponding domains and outcomes, and with maximal scores and threshold values for fails, respectively

Gold standard – neuropsychological test battery		
Domain	Neuropsychological test	Outcome
Language	Semantic Verbal Fluency Test – animal naming (W. I. M. Van Der Elst et al., 2006)	Total correct
Visuospatial ability	Rey–Osterrieth Complex Figure (ROCF) – copy trial (Rey, 1941)	Copy score
Information processing speed	Stroop Color-Word Test (W. Van der Elst et al., 2006)	Time Cards I and II
Executive function	Stroop Color-Word Test	Time Card III
	Brixton Spatial Anticipation Test (BSAT) (Van Den Berg et al., 2009)	Total errors
Attention and working memory	Digit Span WAIS-III (Wechsler, 1997)	Total score forward and backward
Episodic memory	Rey Auditory Verbal Learning Test (RAVLT) (Van der Elst et al., 2005)	Total correct trial 1–3, delayed recall
Affect	Emotion Recognition Test (ERT; computer-assisted) (Kessels et al., 2014)	Score per emotion (anger, disgust, sadness, fear, happiness, surprise)

CCAS scale		
Test item	Maximum score (Hoche et al., 2018)	Fail if score ≤
Semantic fluency	26	15
Phonemic fluency	19	9
Category switching	15	9
Digit span forward	8	5
Digit span backward	6	3
Cube draw/copy	15	11
Verbal recall	15	10
Similarities	8	6
Go/no-go	2	0
Affect	6	4

Since clear criteria for the diagnosis of CCAS are lacking, we considered an extensive neuropsychological test battery including CCAS-related cognitive domains as the gold standard, using established cut-off scores to classify an individual as cognitively impaired. This allowed us to include cerebellar patients with and without CCAS, as well as HC. All patients were assessed with the CCAS scale (± 20 min) and the full neuropsychological test battery (± 60 min). Both were administered with at least one week in between to avoid practice, interference, or fatigue effects. All HC were administered the CCAS scale and the MoCA. The MoCA was used to ensure that no cognitive impairments were present; all controls scored ≥26 points (range: 26–30) (Nasreddine et al., 2005). Cognitive assessment was performed in a standardized manner with trained assessors. Background variables included age, sex, education level (Verhage, 1965), disease stage based on ambulatory status (Klockgether et al., 1998), SARA (scale for the assessment and rating of ataxia) score, disease duration/time since stroke, and clinical diagnosis.

Gold standard – neuropsychological test battery

An extensive test battery was used as the gold standard to establish or refute a CCAS diagnosis. This battery consisted of widely used, reliable, and validated neuropsychological tests and included the cognitive domains that are typically compromised in CCAS, as summarized in Table 1. For the majority of tests, raw outcomes were converted into age-, sex-, and education-corrected *z*-scores with the use of normative values from a large Dutch dataset (de Vent et al., 2016). For the Emotion Recognition Test, published normative data were used and converted into age-, education-, and sex-adjusted *z*-scores (Kessels et al., 2014). Impairment of individual test outcomes was defined by a *z*-score below 1.5 standard deviation (SD) from the normative mean. A patient was classified as cognitively impaired when a patient scored three or more test outcomes below 1.5 SD or two or more tests below 2 SD (Fischer et al., 2014). Cognitive performance validity was assessed using the Reliable Digit Span measure as an embedded performance validity measure (Webber & Soble, 2018).

The CCAS scale

The CCAS scale was developed by Hoche et al. (Hoche et al., 2018) and has four parallel versions to attenuate learning effects and facilitate test–retest reliability. Only version 1A has undergone psychometric testing in individuals with cerebellar ataxia, therefore the authorized Dutch translation of version 1A was used in this study (Mariën et al., 2021). A Dutch administration procedure was also developed (see Supplementary Information 1). The CCAS scale consists of 10 scored items, which are also listed in Table 1. All test items have objective scoring criteria, except for the Affect item which entails a subjective rating of neuropsychiatric domains by the assessor, with input from the patient and caregiver. Each item was scored and the total raw score of the scale ranges from 0 to 120, with lower scores reflecting worse cognitive performance. For each test item, there is also a threshold score to determine a pass or fail. According to the evaluation criteria of the developers (Hoche et al., 2018), one failed item is indicative for "possible" CCAS, two fails for "probable" CCAS, and three or more fails for "definite" CCAS.

Dysarthria correction

As an addition to this study, the influence of dysarthria in patients on test results was examined. Dysarthria was quantified using the PATA Rate Task (PRT) for speech rate; patients were instructed to repeat "PA-TA" as quickly as possible in 10 s and the number of correct repetitions was the score. Two trials were performed, and the average of both was calculated. The PRT score was applied to correct for dysarthria on the timed neuropsychological tests (Semantic Verbal Fluency test and the Stroop Color-Word Test) if the PRT score was below the lower limit of normality threshold (Schmitz-

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Hübsch et al., 2008). This limit was calculated using previously published normative values of the PRT mean and SD in controls, resulting in a PRT score of 15.6 (mean – 2 × SD = 28.84 – 2 × 6.6) (Pane et al., 2018; Saccà et al., 2018).

For correction of the Semantic Verbal Fluency test score, the formula previously established by Saccà et al., was used (Saccà et al., 2018). This formula used the PRT to calculate a corrected time in which the test could be performed. Since we performed the PRT after the Fluency test, we subsequently calculated the corrected score per ratio with the corrected time (corrected score $= \frac{\text{corrected time}}{60} \times$ original score).

For correction of the Stroop Color-Word test score, we established a formula based on the formulas provided by Saccà et al. (2018). Correction could only be applied to Stroop cards II and III scores, as the score of card I was used as a reference. The following formula was used: corrected time $= \left(\frac{\text{original time} \times \text{repetition ratio}}{\text{lower limit of normality} \times \text{PRT score}}\right) +$ cognition time). The original time is the uncorrected Stroop score of card II or III. The repetition ratio is the normative mean of card I divided by the normative mean of card II or III. For card II this is 0.752 (43.50/57.87), and for card III this is 0.416 (43.50/104.52) (Schmand et al., 2012). The cognition time is the original time minus the original time, multiplied by the repetition ratio (Saccà et al., 2018).

Statistical analyses

Analyses were performed using IBM SPSS Statistics 27.0 (SPSS, Inc., Chicago, IL, USA). Means with SDs or medians with interquartile ranges were reported, as appropriate. Between-group differences in CCAS scale outcomes were tested using (Quade's) ANCOVA with education as covariate. Cronbach's alpha was calculated to report internal consistency of the scale, considered acceptable if ≥0.7 (Nunnally, 1967). Construct validity was evaluated in terms of item-total correlations. Sensitivity and specificity were calculated to report on validity and accuracy; minimum values for acceptable sensitivity and specificity were 80% and 60%, respectively (Blake et al., 2002). ROC analyses were performed to determine the AUC as a discriminating measure. AUC values <0.7 were considered poor, between 0.7 and 0.8 as acceptable, between 0.8 and 0.9 as good, and between 0.9 and 1.0 as excellent (F. Li & He, 2018). In addition, we explored whether the validity of the CCAS scale could be improved by simple adaptations. We assessed whether deleting test items with many false-positive results would improve the validity. Furthermore, we applied a simple correction for lower education, where we reduced the number of fails by one for patients with Verhage education level ≤ 5 (comparable to ≤ 12 education years). Also, we applied the correction formula controlling for age, sex, and education effects from the recent study by Liu et al. (2024) on our data, to evaluate whether this could improve the validity. Most optimal threshold values for individual CCAS scale items were determined using the Youden index, where most of the focus was on obtaining acceptable specificity (Youden, 1950). The relationship of the CCAS scale with age, sex, education, disease duration/time since stroke, and disease stage was examined using (Spearman or Point-Biserial) correlations, as well as the coherence of the scale with the gold standard. Since this was exploratory, no correction for multiple testing was applied. Statistical significance was set at 0.05 (two-tailed) for all tests. The anonymized datasets used and analyzed during this study are available from the corresponding author on request from a qualified investigator.

Results

Participant characteristics

According to the gold standard, 49 cerebellar patients were classified as having CCAS, and 30 cerebellar patients were classified as not having CCAS, serving as "cerebellar controls" (CC). Patients were heterogeneous in terms of diagnoses; 19 had a cerebellar stroke, and 60 had a degenerative cerebellar ataxia (details are provided in Supplementary Information 2). Additionally, 32 HC were included. Characteristics per group are shown in Table 2. Validity of the outcomes was evaluated by comparing CCAS patients with CC, or with CC and HC combined. Considering these comparisons, age (CCAS *vs.* CC: $p = .590$; CCAS *vs.* CC + HC: $p = .071$) and sex (CCAS *vs.* CC: $p = .172$; CCAS *vs.* CC + HC: $p = .703$) did not significantly differ. Proportions regarding disease stage were also not significantly different (CCAS *vs.* CC: $p = .148$). Education level was slightly, yet significantly different from the CCAS patients in both CC ($p = .039$) and CC + HC ($p < .001$). The average time between the assessment of the gold standard and CCAS scale was 23.5 ± 77.4 days.

Gold standard – test battery performance

The outcomes of the test battery are shown in Figure 1. The cognitive performance of all patients was considered valid as evaluated using the Reliable Digit Span measure as an embedded performance validity test. Small differences were found in most outcomes between CC and CCAS patients, except for the three subtests of the Stroop Color-Word Test. This test was most frequently impaired in CCAS patients and showed the lowest *z*-scores with the most variance. Especially Stroop card III seemed to discriminate well, since none of the CC showed impaired performance, while 59% of the CCAS patients did. All *z*-scores and raw scores per test outcome are provided in Supplementary Information 3.

CCAS scale performance

Outcomes of the CCAS scale per group are listed in Table 3. Between-group differences were assessed with correction for education since this significantly differed between the groups. The number of failed items was significantly higher for CCAS patients than CC ($F(1,76) = 12.38$, $p < .001$) and CC with HC combined ($F(1,108) = 30.07$, $p < .001$). The total score was significantly lower for CCAS patients than CC ($F(1,76) = 16.51$, $p < .001$) and CC with HC combined ($F(1,108) = 38.45$, $p < .001$). The high percentage of failed test items in the control groups for phonemic fluency, category switching, digit span forward, verbal recall, and affect is notable. Cronbach's alpha for reliability was 0.707 considering the ten items of the CCAS scale. Deletion of certain test items would not result in higher alpha scores. However, three test items had low item-total correlations: cube draw/copy ($r = .171$), similarities ($r = .253$), and go/no-go ($r = .196$).

Validity

Sensitivity and specificity were obtained for each of the CCAS scale outcomes; one fail ("possible"), two fails ("probable"), and ≥ three fails ("definite"). Sensitivity was 98%, 88%, and 65% for, respectively, one, two, or ≥ three fails. The specificity when considering only the CC was 10%, 43%, and 67% for one, two, or

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Table 2. Participant characteristics per group

	CCAS patients	Cerebellar controls (CC)	Healthy controls (HC)
N	49	30	32
Age, *y*	56.6 ± 13.2	58.1 ± 9.6	43.0 ± 19.3
Sex, men	23 (47%)	19 (63%)	13 (41%)
Education level (Verhage, 1964)			
– Below 2y of low-level second. education	1 (2%)	0	0
– Finished low-level second. education	6 (12%)	3 (10%)	0
– Finished average-level second. education	24 (49%)	11 (37%)	2 (6%)
– Finished high-level second. education	18 (37%)	11 (37%)	18 (56%)
– University	0	5 (16%)	12 (38%)
Disease stage (Klockgether et al., 1998)			–
– No gait difficulties	5 (10%)	1 (3%)	
– Gait difficulties	29 (59%)	22 (73%)	
– Loss independent gait	9 (19%)	7 (24%)	
– Confinement to wheelchair	6 (12%)	–	
SARA score (n=34)	9.9 ± 6.9 (n=34)	–	–
Disease duration/time since stroke, y	8 (3-16.5)	7.5 (2.75–15)	–
Clinical diagnosis			–
– Degenerative ataxia	35 (71%)	25 (83%)	
– Cerebellar stroke	14 (29%)	5 (17%)	
PATA Rate Task	18.5 ± 6.2[a]	23.6 ± 3.9	–
Montreal Cognitive Assessment (MoCA)	–	–	28.3 ± 1.3

SARA = scale for the assessment and rating of ataxia
[a] *n* = 47.

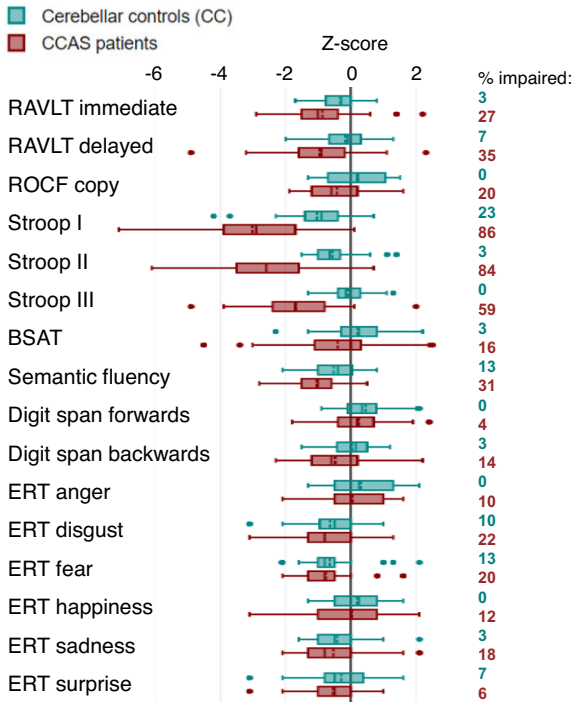

Figure 1. Combined boxplot with *z*-scores of test battery outcomes, including percentages of patients with impairment (<1.5 SD). Boxes represent outcomes within the 1st and 3rd quantile, with solid lines indicating medians and dashed lines indicating means. Whiskers represent the minimum and maximum values (without outliers); outliers are indicated by dots. RAVLT = Rey Auditory Verbal Learning Test; ROCF = Rey–Osterrieth Complex Figure; BSAT = Brixton Spatial Anticipation Test; ERT = Emotion Recognition Test.

≥ three fails, respectively. Specificity was higher when considering both cerebellar and healthy controls: 32%, 68%, and 81% for, respectively, one, two, or ≥ three fails.

The discriminative ability of the CCAS scale as evaluated by ROC analyses is shown in Figure 2. The AUC yielded 0.743 (95% CI: 0.634–0.851) for failed items and 0.762 (95% CI: 0.654–0.869) for the total score when considering only CC. AUC values yielded 0.836 (95% CI: 0.763–0.910) and 0.851 (95% CI: 0.782–0.919), respectively, when considering both cerebellar and healthy controls together.

Optimal threshold values

Sensitivity and specificity for individual test items based on either the original scale's thresholds or the most optimal thresholds determined by the Youden Index are shown in Table 4. Low sensitivities for the majority of test items with original threshold values are improved when Youden threshold values are taken. However, specificity decreases due to the higher Youden threshold values, and their application would lead to more false-positive results and worse overall specificity of the scale (8–44%). Note that the Youden threshold for the similarities and go/no-go items is the maximum score of that item.

Correlations

In our entire sample, outcomes of the CCAS scale were correlated with age (total score ρ = -.328, $p < .001$; failed items $\rho = .290$, $p = .002$) and education (total score $\rho = .439$, $p < .001$; failed items $\rho = -.508$, $p < .001$). No significant correlations were found for sex, disease duration/time since stroke, and disease stage. Test items of the CCAS scale that were comparable to outcomes of the gold standard test battery were also examined, and moderate correlations were found for the semantic fluency tests ($\rho = .637$, $p < .001$) and digit span forward ($\rho = .572$, $p < .001$). Significant correlations were found for the digit span backward ($\rho = .389$, $p < .001$) and delayed recall ($\rho = .372$, $p < .001$). A small correlation was found for the visuospatial items (Rey-Osterrieth Complex Figure copy and Cube draw) with $\rho = .289$, $p = .010$.

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Table 3. Outcomes of the CCAS scale per group

	CCAS patients (n = 49)		Cerebellar controls (CC, *n* = 30)		Healthy controls (HC, *n* = 32)	
Total failed items	3.2 ± 1.6		1.9 ± 1.2		0.7 ± 0.9	
Total score	83.1 ± 11.4		94.2 ± 10.1		105.5 ± 9.2	
Test item:	Score	Failed	Score	Failed	Score	Failed
– Semantic fluency	19.0 ± 5.1	13 (27%)	22.4 ± 3.7	1 (3%)	23.7 ± 3.9	2 (6%)
– Phonemic fluency	7.7 ± 3.1	36 (74%)	10.4 ± 3.8	15 (50%)	13.5 ± 3.4	5 (16%)
– Category switching	9.8 ± 3.5	20 (41%)	11.4 ± 3.2	9 (30%)	13.3 ± 2.2	2 (6%)
– Digit span forward	5.7 ± 0.9	17 (35%)	6.4 ± 1.0	5 (17%)	6.9 ± 1.1	4 (12%)
– Digit span backward	4.3 ± 0.8	6 (12%)	4.6 ± 0.8	2 (7%)	5.3 ± 0.9	2 (6%)
– Cube draw/copy	13.9 ± 1.6	3 (6%)	14.4 ± 1.3	1 (3%)	14.6 ± 1.0	0
– Verbal recall	10.7 ± 3.5	18 (37%)	11.4 ± 3.7	10 (33%)	13.0 ± 2.4	6 (19%)
– Similarities	7.3 ± 0.6	3 (6%)	7.5 ± 0.5	0	7.6 ± 0.5	0
– Go/No-go	1.6 ± 0.6	4 (8%)	1.8 ± 0.4	0	1.7 ± 0.5	1 (3%)
– Affect	3.0 ± 1.4	42 (86%)	4.0 ± 1.7	16 (53%)	6.0 ± 0.2	0
Outcome (fails):						
– No CCAS (0)	1 (2%)		3 (10%)		17 (53%)	
– Possible CCAS (1)	5 (10%)		10 (33%)		12 (38%)	
– Probable CCAS (2)	11 (22%)		7 (24%)		1 (3%)	
– Definite CCAS (≥3)	32 (66%)		10 (33%)		2 (6%)	

Variables are presented as means with standard deviations or frequencies with percentages.

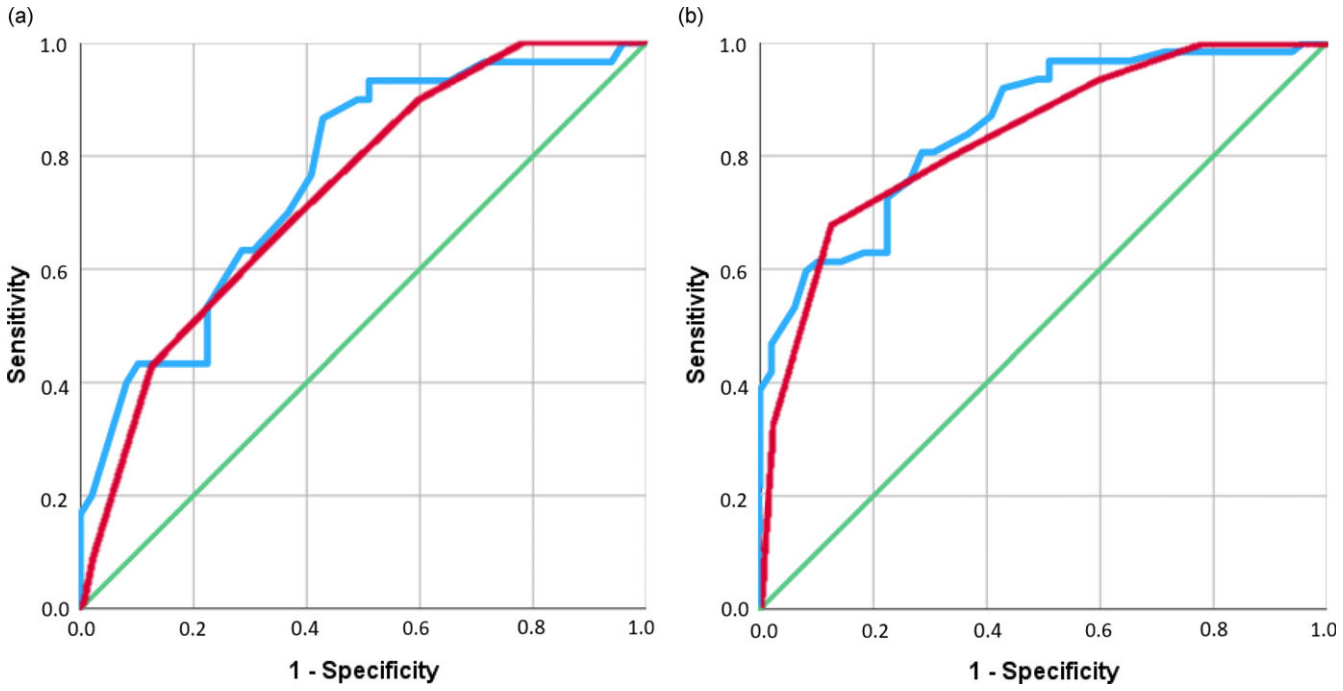

Figure 2. ROC curves for CCAS scale failed items (in red) and total score (in blue), the green line indicates the reference line. **(a)** shows the ROC curve only for cerebellar controls. **(b)** shows the ROC curve for cerebellar and healthy controls combined.

For the verbal fluency items, CCAS scale outcomes were also compared to age-, sex-, and education-corrected Dutch normative data (de Vent et al., 2016). Of the 36 CCAS patients who failed the phonemic fluency item of the CCAS scale, 21 patients were not impaired on this test (z-score < −1.5 SD) according to the normative data. On the other hand, of the 13 patients who failed the semantic fluency item, only two patients were not impaired on this test according to the normative data, while 11 patients with impairments according to the normative data (z-score < −1.5 SD) did not fail the CCAS scale item.

Dysarthria correction

CCAS patients scored significantly lower on the PRT than CC ($p < .001$). The eighteen patients who scored below the lower limit of normality (range: 8–15.5) all had CCAS. For these patients, outcomes of the semantic fluency and Stroop Color-Word Test in the gold standard test battery were corrected, resulting in different group allocations. Six patients (out of 18; 33%) were defined as having CCAS without the correction, and as cerebellar control with the correction. Dysarthria correction did not affect the validity of the scale considerably, since the corrected sensitivity (65–100%) and specificity (11–61%) were similar to

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Table 4. Sensitivity and specificity of CCAS scale test items, with original threshold values and thresholds determined by Youden Index

	CCAS patients and cerebellar controls (CC)		CCAS patients and all controls (CC + HC)	
CCAS scale test item	Original threshold Sens./spec.	Youden threshold Sens./spec.	Original threshold Sens./spec.	Youden threshold Sens./spec.
Semantic fluency	≤15 27%/97%	≤23 78%/60%	≤15 27%/95%	≤23 78%/66%
Phonemic fluency	≤9 74%/50%	≤10 74%/50%	≤9 74%/68%	≤11 82%/63%
Category switching	≤9 41%/70%	≤11 57%/53%	≤9 41%/82%	≤12 65%/60%
Digit span forward	≤5 35%/83%	≤7 82%/37%	≤5 35%/86%	≤7 82%/50%
Digit span backward	≤3 12%/93%	≤5 59%/53%	≤3 12%/94%	≤5 59%/69%
Cube draw/copy	≤11 6%/97%	≤14 33%/83%	≤11 6%/98%	≤14 33%/86%
Verbal recall	≤10 37%/67%	≤12 53%/57%	≤10 37%/74%	≤13 67%/58%
Similarities	≤6 6%/100% ≤0 8%/100%	≤8 61%/50% ≤2 35%/77%	≤6 6%/100% ≤0 8%/98%	≤8 61%/57% ≤2 35%/74%
Affect	≤4 86%/47%	≤5 86%/47%	≤4 86%/74%	≤5 86%/74%
Total score	–	≤86 57%/87%	–	≤91 71%/81%
Total failed items	–	≤3 65 %/67%	–	≤2 88%/68%

those before correction. PRT performance was found to be correlated with both the total score ($\rho = .401$, $p < .001$) and the number of fails ($\rho = -.373$, $p < .001$) of the CCAS scale. Correlations between the PRT and the individual test items of the scale were significant for semantic ($\rho = .305$, $p = .007$) and phonemic ($\rho = .257$, $p = .024$) fluency, category switching ($\rho = .262$, $p = .021$), and digit span forward ($\rho = .302$, $p = .008$).

Adaptations

The influence of simple adaptations on the validity of the CCAS scale was explored. When deleting test items with many false-positive results, such as the phonemic fluency item, the specificity increased, but not sufficiently. The specificity when considering only the CC was 20%, 53%, and 80% for one, two, or ≥ three fails, respectively. When applying a simple correction for lower education, where we reduced the number of fails by one for patients with Verhage education level ≤ 5 (comparable to ≤ 12 education years), the specificity slightly increased, but the diagnostic accuracy remained insufficient. The specificity when considering only the CC was 27%, 50%, and 80% for one, two, or ≥ three fails, respectively. Applying the age, sex, and education correction as established by Liu et al. (2024) resulted in a sensitivity of 86% and a specificity of 37% when considering only CC. The specificity was 52% when also HC were included.

Discussion

This was the first validation study of the CCAS scale that explicitly distinguished between patients with and without a proven CCAS, as defined by a gold standard neuropsychological examination. Patients with CCAS scored significantly worse on the scale compared to CC and all controls combined. The sensitivity of the scale was acceptable (65–98%), but specificity was insufficient, especially when only considering CC (10–67%). ROC analyses showed acceptable discriminative ability at the group level, but validity at the individual level is poor due to the frequent presence of false-positive outcomes in both control groups. Outcomes of the scale were correlated with education and age and should be taken into account for improving the validity of the CCAS scale.

Insufficient specificity due to high false-positive rates of the CCAS scale was found, leading to overdiagnosis of CCAS as a clinical syndrome. This is in line with other recent studies, that challenge the evaluation criteria of Hoche et al. (2018) for the diagnosis of CCAS (Alan et al., 2024; Maas et al., 2021; Selvadurai et al., 2024; Thieme et al., 2022). High false-positive rates are probably caused by a variety of performance among different age groups and education levels. We found that control participants without cognitive impairments commonly failed test items that are age- and education-sensitive, such as verbal fluency, category switching, and verbal recall. CCAS scale outcomes were significantly correlated with age and education level, but these aspects are currently not taken into account in the interpretation of outcomes. Moreover, the discrepancies we found between verbal fluency outcomes of the CCAS scale and outcomes corrected with normative data further illustrate the importance of age and education level in the interpretation of outcomes. The fact that education and age are essential factors to consider has been stressed before by Thieme et al., who observed more false-positive outcomes in control participants with lower education (Thieme et al., 2021). The authors of the CCAS scale replied that a correction for education could indeed be required when testing populations with lower educational levels (Schmahmann et al., 2021). In accordance with the first comment about the importance of age and education by Thieme et al. (2021) and our results, other studies also found a relationship with education and/or age, suggesting that a correction for both factors could improve the scale's properties (Rodríguez-Labrada et al., 2021; Thieme et al., 2022).

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Although the CCAS scale has insufficient validity in its current form, we explored whether its usefulness for clinical and scientific purposes would be improved by adaptations. Deleting test items with many false-positive results did not improve the validity or reliability. Also, applying a simple correction for lower education did not improve the diagnostic accuracy. Another approach was to adjust the threshold values. Adjusting thresholds per test item is not recommended, as the thresholds proposed by the Youden Index did not increase the validity of the scale. Application of a threshold for the total score or number of failed items was also considered but did not yield acceptable sensitivity in the group of cerebellar patients. Thus, a more detailed adjustment for education and age will be required. The recent study by Liu et al. (2024) employed a correction formula that controls for age, sex, and education effects, aiming for an improved evaluation of the CCAS scale outcomes. However, when applying this correction to the data in our cohort, the sensitivity (86%) and specificity (CC: 37%, CC + HC: 52%) unfortunately remained insufficient (Liu et al., 2024). An alternative is that normative values should be established in larger samples, similar to what has been done previously for the MoCA (Kessels et al., 2022). Education- and/or age-stratified reference values will have to be established before the scale can be recommended for reliable use in daily clinical practice.

CCAS scale items assumed to reflect the same cognitive processes as the test battery outcomes were not strongly correlated. For some items, like semantic fluency, we would expect a strong correlation because this item was exactly the same as in the gold standard test battery. Therefore, we examined whether the scores were systematically higher at the second assessment to detect a learning or practice effect, but this turned out not to be the case. Furthermore, we found that the Stroop Color-Word Test was most frequently impaired in patients and discriminated best between patients with CCAS and those without, even after dysarthria correction. A similar item is not included in the CCAS scale and could be considered.

All patients who scored below the lower limit of the normality threshold of the PRT, indicative of dysarthria, also had CCAS. Although dysarthria did not seem to affect the validity of the CCAS scale, 33% of patients with dysarthria (6/18) were classified as having CCAS without dysarthria correction, but as cerebellar control with the correction. PRT performance was correlated with the total CCAS scale score, but also with the fluency items, category switching, and digit span forward, which were commonly failed in the cerebellar control group. A previous study on Friedreich's ataxia found similar significant relationships between measures of speech and verbal test items (Corben et al., 2024). Articulation speed as a potential confounding factor has been mentioned before, as well as the suggestion to correct this in the timed items of the CCAS scale (Chirino-Pérez et al., 2021). Failure on these items may reflect slower speech production rather than cognitive deficits in verbal fluency, although deficits may still be evident when dysarthria is considered (Bolceková et al., 2017; Cocozza et al., 2018; Y. Li et al., 2023; Stoodley & Schmahmann, 2009). This nevertheless illustrates that dysarthria, a common symptom in cerebellar disorders, may influence timed neuropsychological test outcomes, and should be taken into account to prevent misclassification (Paap et al., 2016). Hence, caution is warranted also in the CCAS scale, of which 50% of the total score is determined by dysarthria-sensitive items.

The use of an extensive neuropsychological test battery consisting of reliable and validated tests as the gold standard to substantiate the CCAS diagnosis of participants is a strength of this study. This also allowed us to include a control group of cerebellar patients without CCAS and increased the relevance for clinical practice. Slightly disadvantageous outcomes were observed when only considering the CC compared to all controls taken together, which illustrates the requirement to include a cerebellar control group in validity studies. The inclusion of an etiologically heterogeneous group of cerebellar patients has increased the external validity of our study. Another strength is that we explored the influence of dysarthria and took this into account by correcting test outcomes. Several limitations of our study should be mentioned. First, selection bias may have occurred due to our recruitment approach. Second, we could not provide a more detailed description of disease severity in our cohort. We had information on the ataxia disease stage for all patients, while recent SARA scores were available for only a subset. Also, we purposely included a mixed-etiology ataxia cohort, as this reflects clinical practice. The CCAS scale was specifically developed to serve the purpose as a cognitive screen for any cerebellar patient. However, this "one size fits all" approach can be criticized, and the inclusion of a larger, more etiologically homogeneous ataxia subsample may have provided different results and a more fine-grained cognitive profile for that specific etiology. Furthermore, the HC group was significantly higher educated than both patient groups. However, had the education level been more similar to that of the patients, this would probably have resulted in even more false-positive outcomes in the controls. Finally, we are unable to draw conclusions about test–retest reliability of the scale, because we have no data about the parallel versions.

The need for a validated screener to detect CCAS is high, and there also have been attempts to detect CCAS with (a brief combination of) other tests (Bolceková et al., 2017; Starowicz-Filip et al., 2022). The current scale does not contain an item about social cognition (e.g. an item on mentalizing), which appears to be commonly affected in cerebellar patients, and the Affect item is rather brief and uses observer ratings rather than a performance task (Van Overwalle et al., 2019). Since the variety in neuropsychiatric symptoms is large, it may be better to capture them on a separate scale. For instance, the Cerebellar Impulsivity-Compulsivity Assessment Scale and the Cerebellar Neuropsychiatric Rating Scale have been developed for specific use in cerebellar patients (Karamazovova et al., 2023; Lin et al., 2023; Shao et al., 2024). Future research will have to focus on establishing more extensive normative data for CCAS, thereby explicitly taking education and age into account (Thieme et al., 2021). Subsequently, longitudinal studies including the parallel versions should be performed to assess test–retest reliability and gain insight into whether the scale is suitable to monitor changes in cognitive function over time.

Conclusion

In conclusion, we recommend caution when using the CCAS scale in its current form in clinical practice due to its poor specificity. Previous studies have raised similar concerns, but the scale is already being used as a diagnostic tool and endorsed as a "promising biomarker" (Abderrakib et al., 2022; Bolzan et al., 2024; Selvadurai et al., 2024). We argue that the scale is not yet suitable

https://doi.org/10.1017/S1355617725101033 Published online by Cambridge University Press

for diagnostic purposes in clinical practice and that it may still be recommended to perform an extensive neuropsychological assessment in cerebellar patients with cognitive complaints.

Supplementary material. For supplementary material accompanying this paper visit https://doi.org/10.1017/S1355617725101033.

Acknowledgments. The authors thank Allan Pieterse, Hanneke van Duijnhoven, Sirwan Darweesh, Ilse Willemse, Teije van Prooije, and Lotte van de Venis for their help in the recruitment of cerebellar patients in the control group. We also thank Frank van Overwalle for his contribution to the translation of the CCAS scale to Dutch/Flemish. We are grateful to all participants who participated in this study.

Funding statement. Financial support was provided by a grant from the Hersenstichting Nederland (grant number DR-2019-00313).

Competing interests. The authors have no conflicts of interest to disclose.

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