



# ALS CBS ALS Cognitive Behavioral Screen

## MANUAL

### Amyotrophic Lateral Sclerosis Cognitive Behavioral Screen (ALS-CBS)

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#### **Background and General Description**

The ALS Cognitive Behavioral Screen (ALS-CBS) is a brief measure of cognition and behavior in patients with Amyotrophic Lateral Sclerosis (ALS). Up to half of patients with ALS may develop cognitive impairment during the course of the disease [1,2], and anywhere from 3-41% of ALS meet criteria for frontotemporal dementia (FTD) [1,2]. Patients with ALS who develop these types of impairments have significantly shorter survival compared to other ALS patients and are more likely to be noncompliant with interventions [3,4]. Despite this, no standardized screening measure exists to identify ALS patients who may be cognitively or behaviorally impaired, and no tools are available for the longitudinal study of impairment specific to ALS.



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Standard neuropsychological test batteries are time consuming, and patients with ALS are typically unable to complete them due to physical weakness, paralysis, loss of speech, or severe respiratory compromise. Moreover, most ALS centers and neurology clinics do not have neuropsychologists trained to complete detailed testing with this unique patient population. A validated, brief, and practical measure that could be administered by a variety of clinicians would fill the current gap in assessment.

The ALS CBS is composed of two sections: cognitive and behavioral. The cognitive section includes commonly used elements of standard testing batteries, consisting of 8 tasks. It can be administered by a physician or other clinical care staff and takes approximately 5 minutes to complete. The behavioral section is composed of questions sensitive to organic brain changes. It consists of a set of questions that compare changes in personality and behavior since the onset of ALS, as well as yes/no questions about mood, pseudobulbar affect, and fatigue. It is completed by a caregiver, family member, or other informant during the same time that the patient completes the cognitive portion. This questionnaire typically takes about 2 minutes.

In addition to its clinical utility, the ALS CBS may serve as a standardized and reliable research measure. It is intended for use in longitudinal assessment in order to help answer a longstanding question about whether cognitive or behavioral impairments progress in ALS patients. More specifically, the screen may provide information as to whether patients with ALS eventually develop FTD. The ALS CBS was developed to minimize demands on speech and motor capacities, so that patients can be tested during later stages of disease. Most items can be responded to using augmentative communication devices, or mouthing.

### **Development**

The construction of the ALS CBS was completed in several stages. First, literature was reviewed covering topics of cognitive impairment in ALS, behavioral abnormalities in neurologic diseases, frontal lobe dysfunction, frontal lobes tests, and FTD. Existing neurocognitive screens used in neurologic diseases like dementia were reviewed. Studies evaluating the sensitivity and specificity of various neuropsychological measures in ALS patients were reviewed [1,2,5] and appropriate items were given high preference for inclusion. Consultation with a number of neurologists and neuropsychological experts in the field of ALS and motor neuron disease (MND) occurred. A meeting of several clinicians at the 2006 meeting of the Western ALS (WALS) Research Group delineated the basic structure of the screen and recommended components for inclusion.

Most cognitive items included in the ALS CBS were chosen based on high diagnostic value for frontally-mediated cognitive impairments unique to ALS. Items were also selected if they were easy for clinicians to learn, particularly for those without prior experience with neuropsychological screening tools. Items were preferentially selected if they were brief and not



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highly correlated with education. Items requiring minimal motor and speech involvement were also given relative preference.

Behavioral questions were chosen based on diagnostic criteria for FTD available at the time of initial development [6] as well as initial behavioral research in the field of ALS [7,8]. Existing behavioral screens are not tailored to assess patients with ALS, who suffer from prominent and progressive physical and respiratory disability. As a result, items developed for this measure were worded to minimize the chance of endorsement based simply on disease progression. For example, apathy items were worded to assess attitudes, interest, and intellectual engagement rather than physical activity, which naturally decreases along the disease course.

Yes/No items were included on the behavioral section to track for possible mood disorders, pseudobulbar affect, and fatigue. These were intended to serve as simple screens and research data points and not as a means of diagnosis. Such items were included based on evidence that mood, emotional lability and fatigue can correlate with cognitive and behavioral changes [9-11].

An initial draft of the ALS CBS was administered to 150 consecutive ALS patients at two multidisciplinary ALS centers. All screens were reviewed by the author for accuracy and consistency. The mean cognitive score among all ALS patients was 16.3 (2.85) (total possible score: 20) versus 18.3 (1.0) in controls.

The initial draft of the ALS CBS was also administered to 24 ALS patients from the ALSA Clinic at Baylor College of Medicine with confirmed diagnoses based on cognitive and behavioral data (M=15.3; SD=4.12; 9 unimpaired, 3 cognitively impaired, 7 behaviorally impaired, and 5 FTD) [12]. A one-way analysis of variance revealed a significant difference between diagnostic classifications on the total score of the cognitive portion of the screen ( $p=0.02$ ). The cognitive screen distinguished significantly between the cognitive classifications as scores for ALS patients diagnosed with FTD were significantly lower than both the unimpaired ( $p<0.01$ ) and the cognitively impaired ( $p=0.04$ ) groups; a significant difference between FTD patients and those classified as behaviorally impaired was not found ( $p=0.17$ ). Additionally, a subset of 11 patients received both the cognitive screen (M = 15.8; SD=3.60) and a comprehensive neuropsychological battery within 9 months of each other. The neuropsychological battery assessed multiple domains including memory, visuospatial skills, language, behavior, and executive function. The validation of the cognitive section of the ALS CBS was investigated in this subset of patients with a K-means cluster analysis that resulted in a 91% correct identification of cognitively impaired patients.

York and colleagues [12] also examined the behavior portion of the screen in 24 ALS patients whose caregivers completed both the behavioral portion of the screen (M=19.2; SD=3.64) and the full Frontal Systems Behavior Scale (FrSBe) (M T-Score=62.3; SD=16.8). The behavioral



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screen significantly correlated with the FrSBe Total T score ( $p=0.01$ ), indicating a strong association between the behavioral screen (ALS CBS) and the standardized behavioral measure. A one-way analysis of variance revealed a significant difference between the behaviorally impaired and unimpaired diagnostic classifications with behaviorally impaired ALS patients (ALSbi) performing worse than their unimpaired counterparts ( $p=0.01$ ). However, the behavioral screen did not accurately classify these patients into their respective categories (67%) based on K-means cluster analysis.

### Current Version

Based on these results and feedback from experts in the field, revisions were made to the screen. More extensive instructions were added to the cognitive section to increase inter-rater reliability. For the behavioral questionnaire, additional items were included and the Likert scale was broadened in efforts to enhance the diagnostic accuracy. The initial version of the ALS CBS included parallel forms with similar but not identical cognitive items in order to minimize practice effects over time. However, analysis of initial data suggested that minimal practice effects occurred, even when using the same screen version over three month intervals. As a result, the second version of the ALS CBS includes only one version.

Results of the initial validation study were published in 2010 [13]. Initial analyses of data from 112 ALS patients suggest that ALS CBS™ cognitive scores were correlated with education ( $p=0.01$ ), the ALS CBS behavior score ( $p=0.016$ ) and forced vital capacity (FVC) ( $p=0.04$ ). Behavioral scores were correlated with the cognitive score of the ALS-CBS™ and trended towards a significant correlation with FVC ( $p=0.06$ ). The correlation between standardized change scores (Total Current T score- Total Premorbid T score) from the Frontal System Behavioral Scale (FrSBe) Family Rating Form and the ALS-CBS behavioral score was significant ( $p=0.0026$ ).

Thirty-one ALS patients completed both the ALS-CBS and a neuropsychological assessment battery. These patients had an average age of 56 years, mean education of 14.5 years, an average FVC of 84% and average ALS FRS-R score of 34/48. The majority of patients were limb-onset (66%). Patients were separated into diagnostic classifications of cognitively and behaviourally normal (N=14), cognitively impaired (ALS<sub>ci</sub>; N=8), behaviourally impaired (ALS<sub>bi</sub>; N=6) and ALS patients meeting criteria for FTD (ALS-FTD; N=6) based on results of comprehensive neuropsychological testing.

No differences existed between the ALS CBS scores of normal controls and ALS patients who were characterized as cognitively normal based on neuropsychological testing ( $p=0.355$ ). A significant difference was present on cognitive scores between ALS patients with and without ALS-FTD ( $p=0.0005$ ) (Table 1).

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Table 1: Means and standard deviations of ALS-CBS scores of the initial validation cohort (N=31)

Cohort	Cognitive Score Mean (SD) Total score: 20	Behavior Score Mean (SD) Total score: 45
Normal controls (non-ALS) (N=15)	18.8 (1.3)	42.3 (2.1)
ALS cognitively & behaviourally normal (N=14,10)†	17.7 (1.9)	40.3 (3.8)
ALS cognitive impairment (ALSci) (N=8,5)	16.1 (1.8)	40.8 (4.4)
ALS behavioural impairment (ALSbi) (N=6,4)	16.0 (3.2)	30.2 (12.1)
ALS-FTD (N=6)	3.67 (3.44)**	22.8 (10.6)*

†First number is sample size for cognitive test, second number is sample size for behavioural test. Some patients were included in both the ALSci and ALSbi groups depending on which diagnostic criteria they met.

\*\*p=0.0005 (FTD vs ALS cognitively normal)

\*p=0.005 (FTD vs ALS behaviourally normal)

### Comparison of screen scores across diagnostic groups

Mean cognitive scores for the ALS normal, ALSci, ALSbi and ALSbici groups were not statistically different ( $p=0.35$  by Kruskal-Wallis test), but the number of patients in each group was small. We did find a significant decrease in both total cognitive and total behaviour scores with advancing degrees of cognitive impairment. Linear regression of total cognitive and behavioral scores by diagnostic category (separated by controls, ALS normal, ALS impaired, and ALS-FTD) was significant ( $p<0.001$ ) for each regression.

In direct group comparisons, we found differences in cognitive scores between ALS-normal (N=14) and ALS-FTD patients (N=6) ( $p=0.0005$ ). To determine if non-FTD patients differed from normal controls, we collapsed the remaining four diagnostic groups into a single category (ALS, non-FTD) and found that cognitive scores differed significantly from control subjects ( $p=0.008$ , Mann-Whitney test). ALS, non-FTD also had higher cognitive scores than ALS-FTD ( $p=0.001$ , Mann-Whitney test). Finally, we grouped ALS patients with any cognitive or behavioral impairment (ALS-impaired, N=10) and found that mean cognitive scores were lower than ALS-normal, but the difference did not reach significance ( $p=0.15$ ). ALS-impaired did differ significantly from the ALS-FTD group ( $p=0.001$ ).

ALS-CBS behavioral scores did not differ among ALS subgroups without FTD ( $p=0.16$  by Kruskal-Wallis test). Mean behavioral scores were again collapsed into one diagnostic group (ALS, non-FTD). Behavioral score differences for the ALS, non-FTD group and controls did not reach significance ( $p=0.14$  by Mann-Whitney test). Behavioral scores were marginally greater for the ALS, non-FTD group than for the ALS-FTD cohort ( $p=0.09$  by Mann-Whitney test). Behavioral scores were significantly different between ALS-normal, ALS impaired and ALS-FTD ( $p=0.022$  by Kruskal-Wallis test). The difference between ALS-normal and ALS-FTD was significant ( $p=0.005$ ).



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The behavioral section was administered to a group of ALS caregivers online via the Patients Like Me™ website. Initial results of split-half reliability from 70 caregiver responses were promising (Guttman = 0.847). Cronbach's Alpha of 0.916 suggested that none of the items need to be removed [Wicks, personal communication]. Tukey's test of additivity was not violated, suggesting that a valid total score could be obtained by summing all item scores. Test-retest reliability over a one-week period was strong (0.901).

### **Interpretation of Results**

This screen is intended to identify patients in need of further assessment and should not be used as a substitute for standardized testing or formal assessment. A diagnosis of FTD or other dementia should not be given solely on the basis of this screen. In a research setting, the screen is ideally meant to help categorize patients into groups based on scores.

Screen scores should be interpreted with caution in patients with advanced age (>80) or other risk factors such as significant vascular disease, head injury, or major depression. For older patients, a MMSE or other screen should be considered to rule out Alzheimer's disease or similar dementias. Cognitive CBS scores correlate with education, and therefore results for patients with limited education need to be interpreted with caution. Given the strong correlation between cognitive scores and FVC, consideration of reversible causes of cognitive impairment need to be considered for low scores, and respiratory staff should be alerted to low screen scores which may indicate altered respiratory status. Performance of patients whose primary language is not English should be interpreted with caution.

Cognitive scores: Optimal cut points for cognition and behavioral sections were identified which best differentiated between ALS-FTD and the remainder of the cohort [13]. For the cognitive section, a cutoff of equal to or below 10 had 100% sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) for identifying FTD. Thus, the cutoff of 10 for cognition achieved 100% accuracy.

We further assessed the optimal cutoff that could differentiate between ALS-normal and patients with any cognitive deficit (ALSci, ALSbici, FTD). For cognition, a cutoff score greater than or equal to 17 had 86% NPV and 71% specificity to exclude cognitive impairment, while scores below 17 had 85% sensitivity and 69% PPV that there would be some cognitive impairment on a full battery. The cognitive cut-off of 17 achieved 77% accuracy for detecting any cognitive impairment.

Behavioral scores: A cutoff equal to or less than 32 gave 88% sensitivity, 80% specificity, 94% PPV and 67% NPV. Overall, the cutoff of 32 for behavior achieved 86% accuracy for correctly classifying ALS patients with FTD. A score above 36 has a specificity of 86% and NPV of 92% for predicting that no behavioral impairment would be endorsed on comprehensive evaluation, while a score equal to or less than 36 has 90% sensitivity and 82% PPV for



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predicting behavioral impairment. The cut-off of 36 for behavioral score achieved 86% accuracy for detecting any behavioral impairment (either ALSbi or ALS-FTD).

The mean score for ALS-FTD patients was 22.8 (standard deviation 10.6). The most common behavioral changes endorsed by caregivers of ALS-FTD patients included decreased emotional responsiveness, withdrawal without sadness, confusion or distraction, and decreased awareness/denial of problems and changes.

For ALS patients scoring in the ranges which raise suspicion of FTD, further evaluation is required. Ideally, this would be completed by a diagnostic interview which is supplemented by standardized behavioral measures including but not limited to the Cambridge Behavioral Inventory-Revised [14], Frontal Behavior Inventory [15], or the Frontal Systems Behavioral Scale [16].

### **Future Validation & Revision**

Further validation against the gold standard of neuropsychological testing is ongoing. Examination of possible correlations between ALS-CBS scores and clinical features including region of disease onset, distribution of weakness at the time of testing, distribution of upper motor neuron signs, bulbar severity, respiratory functioning, and duration of disease may enhance our understanding of whether certain phenotypes are at greater relative risk for extra-motor impairments. The correlation between cognitive screen scores and respiratory function also warrants further research. Whether ALS CBS scores correlate with neuroanatomical abnormalities is not known.

The measure has been translated in other languages and validation studies in alternate languages may provide further information about the screen's relative utility. Future versions of the screen may be amended based on additional validation studies, evolving neuropsychological research in ALS, and revised diagnostic criteria for behavioral variant FTD [17].

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(inadvertently overlooked) individuals who have a common goal of accurate assessment and effective treatment of patients with ALS.

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### INSTRUCTIONS

#### **Purpose of Screen**

This screen was developed to identify ALS patients at risk for cognitive and/or behavioral impairment. It is not meant to replace neuropsychological assessment, and should not be used to diagnose ALS-Frontotemporal Dementia (FTD), ALS-cognitive impairment (ALSci) or ALS-behavioral impairment (ALSbi). Patients may be informed that the screen assesses abilities like attention and concentration, which can be affected by many variables such as poor sleep, respiratory impairment, mood, medications, or the effects of the disease on the brain. Completing the screen can help clinicians track cognitive and behavioral functioning over time and consider alterations in treatment, as needed.

#### **Testing Environment**

A quiet exam room free of interruption is ideal. Testing the patient alone is preferable. However if others are in the room, politely ask them not to assist the patient with his/her responses. For all tasks, gentle and positive encouragement can be provided to the patient if they want to stop in the middle of a difficult task. However, no assistance with answering can be provided by the examiner or observer.

The behavioral questionnaire (second page of the screen) should be provided to the caregiver at the start of the examination to minimize caregiver involvement in the cognitive testing.

#### **Verbal versus Written Format**

This screen can be completed either orally or in writing. The clinician administering the screen should use their best judgment to determine which format to use. It is recommended that bulbar patients, who may lose their speech more rapidly, use the written format across serial assessments rather than switching from verbal to written midway through the disease course. This will allow for a more reliable assessment of change over time. For those writing responses, please provide paper or eraser-board, etc. For patients who cannot speak or write, mouthing responses or using an assistive communication device is appropriate.

#### **Administration Time & Frequency**

The total time for screening is approximately five minutes. More time may be required for patients who are markedly fatigued or who use a communication device. Typically, patients do not need to complete the screen more frequently than every three months. Screening at least once yearly could be considered at minimum.



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### **COGNITIVE SECTION**

Each of the 4 sub-sections described below has a minimum score of 0 and maximum score of 5. The total score for the cognitive section ranges from 0-20, with 20 being a perfect score. To calculate the score for this section, simply sum the scores for the 4 different subsections described below.

#### **ATTENTION** (total score: 5)

- a. **Commands**: Recite the instructions on the form and ask the patient to wait until the entire command is read before beginning their movement. If the patient cannot point, suggest that they respond by using a limb or their eyes. Minor alterations of instructions are allowed, as this task is meant to assess attention and sequencing of 2 and 3-step tasks. Do not repeat the instructions, or if you do, score a 0 but allow the patient to complete the task by repeating the command. The only exception to this is if the patient requires repetition due to hearing impairment.

Errors include any problem with sequencing, leaving out a step in the command, or executing the wrong action. For example, for command #1, if the patient points to their right instead of their left, that would be considered an error. For command #2, if the patient touches their shoulder, then makes a fist and points with the fist to the floor, this would be counted as an error. An exception to this may be if the patient cannot easily move their hand, and an attempt to point appears made; if this is the case, the patient would not be considered incorrect. Any errors result in a score of 0 for this subsection of the Attention subtest.

- b. **Mental Addition/Language**: Recite the instructions on the form. If the patient does not have good command of the English language or otherwise needs clarification about what a syllable is, please explain until the patient comprehends the concept. Providing more than one example is allowed until the concept is understood. Read each phrase clearly and slowly. Repetition of each phrase once is allowed, but the 20 second time limit should start before the repetition. Record the patient's response. Any incorrect calculation or a response that is not provided within 20 seconds after the initial recitation of the sentence is considered an error. Any error results in a score of 0 for this subsection of the Attention subtest.
- c. **Eye Movements**: Sit directly in front of the patient, about 12-24 inches away, and hold up two fingers approximately 36 inches apart, at patient's eye level. For the saccades task: Wiggle one finger. Ask the patient to look at the finger that moves without moving their head, and then look back at you. The examiner should demonstrate this for the patient to ensure that they understand the task, and then have the patient execute 2-3 practice trials before starting. Randomly move either left or right finger, wait for the patient to respond

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with their eyes, and pause for 1-2 seconds between each trial. Complete a total of 8 trials. If all 8 are correct, the score is 1; any error results in a 0 score.

*Saccade Instructions: I am going to hold my fingers up. Please keep your head straight and look at me. When I wiggle a finger, I want you to look at that finger and then look back at me (examiner should execute this eye moment themselves to demonstrate). Look at my finger by moving your eyes only, trying to keep your head still. Each time I wiggle a finger, look at it and then back to me. (Do 2-3 trials with the patient as practice) We will do that a few times. Ready? (Do 8 random trials, pause for 1-2 seconds between each trial).*

Antisaccades: Explain to the patient that he/she will now do something different. Demonstrate the task by wiggling one finger, and instruct the patient to keep their head still and to look directly at the opposite finger without looking at the finger that moves first. Once the eye movement is complete, have the patient look back at you. Complete 2-3 practice trials before starting. Record the number correct out of 8, alternating fingers randomly. Reiteration of instructions is acceptable. 8/8 correct=score of 2, 7/8 correct=score of 1, and 2 or more errors results in a score of 0.

*Antisaccade Instructions: Good, next I am going to wiggle a finger again, but this time, I want you to look AWAY from the finger that moves. For example, if I move this finger (wiggle one) then I want you to look at the other finger, not the one that moves, ok? (Examiner should demonstrate for patient) Let's try it (do 2-3 trials). Just like before, try to keep your head still and just move your eyes. After each one, look back at me. Ready? (Do 8 random trials, pause for 1-2 seconds between each trial).*

Scoring errors: any incorrect gaze, even if the patient immediately self-corrects. An error is also counted if a patient attempts to anticipate your move by looking before you wiggle a finger. As an untimed test, a delay in eye movements is not considered an error unless the delay seems notable (i.e. >10 seconds).

Examiners may want to copy the instructions above and print them on the back of the cognitive section of the screen for reference, or keep a copy in examination rooms along with the rules for verbal fluency (see below). Saccade instructions are included with the ALS CBS attachment (page 3).

### CONCENTRATION (total score: 5)

Instructions: *I am going to say some numbers. After I say them, I want you to repeat (or write) them to me backwards, or in reverse order. For example, if I say 3-6, you would say (write) 6-3. Understand?* If the patient is writing responses, ask them not to write anything until you are finished stating the span. Do not allow them to write the span forwards and then backwards.



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Errors: If the patient repeats the span without reversing the numbers on the first trial, correct them, reiterate the instructions, count #1 as an error, and continue to #2. Any other errors are left uncorrected and counted as errors. If the trial is incorrect but the patient then corrects themselves spontaneously, give them credit for a correct response. Prompting or repeating of spans is not allowed. Discontinue after two consecutive errors. The score is the maximum span correct, even if patient can do a span only once. For example, if a patient completes 8-7-2 (span=3), but is incorrect with 5-8-1 (span=3), 7-8-6-4 (span=4) and 2-5-9-3 (span=4), their score is 3.

### TRACKING/MONITORING (total score: 5)

All of these tasks are untimed.

- a. Months: Ask the patient to say or write the months of the year backwards, starting with December. If the patient writes their responses, do not allow them to write the months forwards first as a reference. If possible, cover up their written responses as they are written. Typically, patients who write responses may write the first letter(s) of each month instead of writing the whole word for each. Mark any uncorrected errors. Do not tell patient when an error is made. Errors include uncorrected responses out of sequence, repetitions, intrusions, or omissions. Circle omissions and write in other errors on the form. An error-free performance results in 2 points, 1 error results in 1 point, and 2+ errors receives 0 points.
- b. Alphabet: Ask the patient to say/write the alphabet, at their own pace (this is not timed). Mark any uncorrected omissions, repetitions or responses out of sequence as an error. If a patient clearly makes an error due to obvious rushing, do not count it as an error. Ask them to attempt the task again at a slower rate, but if errors are made again, count these. Any errors result in a score of 0.
- c. Alternation Task: Read the instructions on the form, explaining the task. Provide the example of alternating between numbers and letters, by saying 1-A, 2-B, 3-C slowly and clearly. Ask the patient to continue on from there until you tell them to stop. Errors are counted as any mistake in sequencing (ie. 4-D, 5-F) or in losing the task rule (i.e. 4-D, E-5, or 4-D, 5, 6, 7-E). If responses are written, again try to cover up their responses as they are provided. Stop at 13-M. An error-free performance results in 2 points, 1 error results in 1 point, and 2+ errors receives 0 points.



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### INITIATION AND RETRIEVAL (total score: 5)

Read instructions on the form to the patient. You can provide examples to clarify the task rules. For example, *If I give you the letter T, you can say truck or trim or tiny, but you do not want to say "Thomas" or "Texas" or "twenty". Also, try not to say the same word with a different ending, like truck, trucks.* Allow the patient to reference the sheet marked Fluency Rules so they are not required to memorize the task rules.

If the patient writes their responses, provide them a blank sheet of paper, and then transcribe their responses onto the screen form. You may also staple their written responses to the form and enter the score on the sheet. They may use an augmentative communication device, although their typing speed may reduce their total score. If the examiner comprehends the intended word before it is fully typed, then this can be communicated to the patient so they have more time to generate other words.

Tell the patient to begin and then record their responses for 60 seconds. If the patient does not respond within a 15-second time period, you may provide a gentle prompt such as "Keep going". Positive encouragement can be provided without distracting the patient. Record each word stated. If you cannot understand the patient due to dysarthria, write what you can and listen for repetitions of that word.

Scoring:  $\geq 12$  correct words=3 points, 8-11 correct words=2 point,  $< 8$  correct words=1 points, and  $\leq 4$  correct words=0 points for the entire subtest, regardless of rule violations. If patient recites only three words, score entire subtest as 0, but mark the number of rule violations on the form.

Errors include repetitions, rule violations (names of people, places, numbers, or the same word with a different suffix), words starting with a different letter, or non-words. Slang and curse words are not counted as errors. Mark with an X next to any errors or circle them. If  $> 4$  correct words are generated, count and score rule violations. If no errors are made and more than 4 total words are generated, the patient gets an additional 2 points. If 1 error is made, the error score is 1. If 2 or more errors are made, the error score=0.

Total the score for correct words generated (0-3) and the error score (0-2). The maximum score is 5.

### **BEHAVIORAL SECTION**

It is recommended that the behavioral questionnaire be provided to the informant (caregiver, family member) at the start of the examination to minimize the involvement of the informant in the cognitive testing. Instructions are written for the caregiver on the questionnaire.



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## ALS CBS ALS Cognitive Behavioral Screen

This should not replace a diagnostic interview for depression or other mood disorders. If depression is suspected, interpret caregiver behavioral responses with caution. If significant fatigue or respiratory distress is present, also interpret apathy questions with caution. If FTD is suspected, a comprehensive interview supported by additional standardized measures of dementia is required. These can include measures like the Cambridge Behavioral Inventory-Revised (Hodges) ([www.ftdrg.org](http://www.ftdrg.org)), or the Frontal Behavioral Inventory (Kertesz).

### **Review of results**

The screen, in its current form, is designed to provide a general impression to staff and not provide a diagnosis. If a neuropsychologist or other clinician is available who has experience with screening measures, they may provide general feedback to the patient.

Initial results suggest that cognitive scores ranging from 17-20 do not support the presence of clear cognitive impairment. Scores below 16 raise suspicion of cognitive impairment, and this suspicion increases significantly as scores fall below 12. Scores below 10 raise considerable suspicion for ALS-FTD or other dementia and suggest the need for further evaluation. Please see the first section of the Manual for more detailed information about the relative accuracy of different cut off scores.