

- RIG:**
- 1=>yes
- 2=>no
- 3=> unknown

Date of RIG:

____/____/____

M M D D Y Y Y Y

unknown

- Site of onset:**
- 1=>bulbar
- 2=>spinal
- 3=>thoracic/respiratory

Forced Vital capacity at diagnosis: _____ (liters)

_____ (percent of predicted)

SNIP at diagnosis: _____

- Phenotype:**
- 1=>Upper motor neuron predominant ALS
- 2=>Lower motor neuron predominant ALS
- 3=>Flail arms
- 4=>Flail legs
- 5=>Bulbar
- 6=>classical ALS

El Escorial category at diagnosis:

1=>suspected ALS

- 2=>possible ALS
- 3=>possible ALS lab supported
- 4=>probable ALS
- 5=>definite ALS

El Escorial category at a later date (table with several possible entries):

Date:

____/____/____
M M D D Y Y Y Y

- 1=>suspected ALS
- 2=>possible ALS
- 3=>probable ALS lab supported
- 4=>probable ALS
- 5=>definite ALS
- 6=>definite FALS

Change of diagnosis other than ALS/MND: _____

ALSFRS score at diagnosis: _____

NEUROPSYCHOLOGY TESTING**Date of testing:**

M	M	D	D	Y	Y	Y	Y

Based on FAB with additional VFI**1. Similarities (conceptualization)***“In what way are they alike?”*

A banana and an orange (In the event of total failure: “they are not alike” or partial failure: “both have peel,” help the patient by saying: “both a banana and an orange are...”; but credit 0 for the item; do not help the patient for the two following items)

A table and a chair

A tulip, a rose and a daisy

Score (only category responses [fruits, furniture, flowers] are considered correct)

Three correct: 3

Two correct: 2

One correct: 1

None correct: 0

2. Lexical fluency (mental flexibility)

“Say as many words as you can beginning with the letter ‘S,’ any words except surnames or proper nouns.”

If the patient gives no response during the first 5 seconds, say: “for instance, snake.” If the patient pauses 10 seconds, stimulate him by saying: “any word beginning with the letter ‘S.’ The time allowed is 60 seconds.

Score (word repetitions or variations [shoe, shoemaker], surnames, or proper nouns are not counted as correct responses)

More than nine words: 3

Six to nine words: 2

Three to five words: 1

Less than three words: 0

3. Motor series (programming)*“Look carefully at what I’m doing.”*

The examiner, seated in front of the patient, performs alone

three times with his left hand the series of Luria “fist–edge–palm.” “Now, with your right hand do the same series, first with me, then alone.” The examiner performs the series three times with the patient, then says to him/her: “Now, do it on your own.”

Score

Patient performs six correct consecutive series alone: 3

Patient performs at least three correct consecutive series alone: 2

Patient fails alone, but performs three correct consecutive series with the examiner: 1

Patient cannot perform three correct consecutive series even with the examiner: 0

4. Conflicting instructions (sensitivity to interference)

“Tap twice when I tap once.”

To be sure that the patient has understood the instruction, a series of three trials is run: 1-1-1. “Tap once when I tap twice.” To be sure that the patient has understood the instruction, a series of three trials is run: 2-2-2. The examiner performs the following series: 1-1-2-1-2-2-2-1-1-2.

Score

No error: 3

One or two errors: 2

More than two errors: 1

Patient taps like the examiner at least four consecutive times: 0

5. Go–No Go (inhibitory control)

“Tap once when I tap once.”

To be sure that the patient has understood the instruction, a series of three trials is run: 1-1-1. “Do not tap when I tap twice.” To be sure that the patient has understood the instruction, a series of three trials is run: 2-2-2. The examiner performs the following series: 1-1-2-1-2-2-2-1-1-2.

Score

No error: 3

One or two errors: 2

More than two errors: 1

Patient taps like the examiner at least four consecutive times: 0

6. Prehension behavior (environmental autonomy)

“Do not take my hands.”

The examiner is seated in front of the patient. Place the patient’s hands palm up on his/her knees. Without saying anything or looking at the patient, the examiner brings his/her hands close to the patient’s hands and touches the palms of both the patient’s hands, to see if he/she will spontaneously take them. If the patient takes the hands, the examiner will try again after asking him/her:

“Now, do not take my hands.”

Score

Patient does not take the examiner’s hands: 3

Patient hesitates and asks what he/she has to do: 2

Patient takes the hands without hesitation: 1

Patient takes the examiner’s hand even after he/she has been told not to do so: 0

Total FAB SCORE (0-18) :

Verbal fluency Index :

Instructions: “Now, I will give you three minutes to tell me as many words as you can think of starting with the letter 'F'. Names of places and people are not allowed, so no 'France' and no 'Fred'. Avoid using two words that are very similar or are the same word with different endings such as 'friend' and 'friendly'. Ready? ”

- The list of words can be written down by patient or spoken. If spoken the examiner should write down all words generated by patient.
- Modality
Written Spoken
- Document the number of words generated at 3 minutes: _____
- If the words were written, ask patient to copy all he/she has generated and document time taken to copy list of words: _____
- If the words were spoken, ask patient to read the list of words that he/she has generated and document time taken to read the list of words: _____
- **Verbal Fluency Index:**
= $\frac{[240 - \text{time taken to copy/read list ()}]}{\text{Number of words generated ()}}$ = _____

Cognitive status classification

- 1=>Normal
- 2=> executive dysfunction
- 3=>non-executive dysfunction

Type of dementia:

- 1=>Not applicable
- 2=>FTD
- 3=>PPA
- 4=>Semantic dementia
- 5=>Amnestic

GENETIC TESTING

Which ALS genes were tested?:

- 1=>SOD1
- 2=>TARDBP
- 3=>FUS
- 4=>ANG
- 5=>VABP
- 6=>SETX
- 7=>DCTN1
- 8=>other _____

Was a mutation shown in an ALS gene?:

- 1=>SOD1
- 2=>TARDBP
- 3=>FUS
- 4=>ANG
- 5=>VABP
- 6=>SETX
- 7=>DCTN1
- 8=>other _____