Revised ALS Functional Rating Scale (ALSFRS-R)

Training slides
Introduction to ALSFRS-R

Background

This is the primary outcome measure for the trial.

The ALS Functional Rating Scale-Revised (ALSFRS-R) is a 12-item functional scale assessing the ability of the people with motor neurone disease to perform daily activities. The scale can be divided into 4 subgroups:

- fine motor function
- gross motor function
- bulbar function (speech and swallowing)
- respiratory function

Each item is scored from 4 (normal) to 0 (unable). This means that a decline in score over time indicates loss of function and disease progression.
Introduction to ALSFRS-R

Validation

The ALSFRS-R is used widely to assess MND disease progression in clinical practice and in clinical trials.

It has been validated for use by both telephone and by videoconferencing and has good inter and intra-rater reliability.
Introduction to ALSFRS-R

Purpose

The purpose of this presentation and accompanying Working Practice Document (WPD) is to ensure standardisation when using the ALSFRS-R scoring system.

Standardisation and the correct procedure for performing, recording and scoring of the ALSFRS-R is essential for accurate results.
Procedure
Administering the scale

The initial question must be asked as stated, but the person administering the questionnaire should explore the patient’s response further if needed.

As a general rule, “help” means help from a person or a device or appliance. For example a handrail, ankle foot orthosis or walking stick would count as help. The only exception is question 5a where modification of cutlery to make the handles larger is allowed (but counts as slow or clumsy).

If someone has for example fallen and broken their ankle and unable to climb stairs but usually can score must reflect that on the day of testing there is an impairment.

The ALSFRS-R is a scale designed to assess function at home as rated by the patient.

It is a series of 12 questions dealing with aspects of the patient’s daily life, each of which is scored 4 to 0; with 4 being ‘normal’.

The patient should not be prompted in any way, except as described, either by the person administering the scale or by a caregiver.
Procedure

Administering the scale (continued)

If the scale is administered over the telephone and the patient is unable to respond because of significant bulbar impairment, a caregiver should relay the questions and responses.

The only situation in which prompting is permitted is if the patient response is clearly at odds with observation. In that case, the person administering the scale should read out the list of choices.

When ALSFRS-R is being conducted either by teleconference or telephone ask if there is anyone else in the room with the patient and then make it clear that the questions should only be answered by the patient without prompting except as described.

Data should be recorded as required.
1. SPEECH

Ask “How is your speech?”

Score 4: Normal speech process. *Speech is exactly the same as before the onset of MND symptoms.*

Score 3: Detectable speech disturbance. *Refers to any change noticed either by the patient or the carer not attributable to an obvious cause such as new dentures.*

Score 2: Intelligible with repeating. *More than 25% of the time, repeating is necessary for comprehension.*

Score 1: Speech combined with non-vocal communication. *Writing, use of speech synthesizers or similar methods are needed to supplement speech.*

Score 0: Loss of useful speech.
2. SALIVATION

Ask “How is your saliva?”

Score as reported regardless of medication use. Some people have a dry mouth. If that is the only problem, score as normal.

Score 4: Normal. *There is no excess saliva (dry mouth is acceptable as normal).*

Score 3: Slight but definite excess of saliva in mouth; may have night time drooling. *There is an excess of saliva, but usually no need to mop up the saliva with a tissue.*

Score 2: Moderately excessive saliva; may have minimal drooling. *A tissue needs to be used, but less than 25% of the time.*

Score 1: Marked excess of saliva with some drooling. *There is likely to be drooling and a tissue is often, but not always used.*

Score 0: Marked drooling. *Requires a constant use of tissue or handkerchief, or suction.*
3. SWALLOWING

Ask “How is your swallowing?”

Score 4: Normal eating habits. There is no change from before symptom onset; the person should be able to eat any food in typical mouthful sizes or drink liquid without difficulty.

Score 3: Early eating problems – occasional choking. Occasionally food will stick, or cause coughing or choking. Food may need to be cut up small, but is not mashed or liquidised.

Score 2: Dietary consistency changes. Food needs to be mashed or liquidised, drinks may need thickener, or some foods such as steak, dry biscuits or cornflakes are avoided in favour of yoghurts, casseroles or porridge.

Score 1: Needs supplemental tube feeding. Oral intake of food is so difficult that significant weight loss (more than 10%) has occurred and gastrostomy is required to supplement caloric intake regardless of whether one is fitted or not.

Score 0: NPO (Exclusively parenteral or enteral feeding).
4. HANDWRITING

Ask “Are you able to hold a pen?” If the answer is “Yes” then ask “How is your writing?” and explore whether words are legible.

Only score the dominant hand and only score for use of a standard pen of normal size.

Score 4: Normal.

Score 3: Slow or sloppy: all words are legible. Use of large pen grips or other writing aids, or any change in writing compared with before symptom onset.

Score 2: Not all words are legible. Ignore ability to write name or sign when scoring.

Score 1: No words are legible, but can still grip pen. Writing is illegible – signing or writing name legibly does not count if the patient has not written other words except their name or signature recently and therefore cannot answer the question further.

Score 0: Unable to grip pen.
5a. CUTTING FOOD AND HANDLING UTENSILS:

Patients without gastrostomy

If someone has a gastrostomy but it is not the primary method of caloric intake, treat as “without gastrostomy”.

Ask “How are you with cutting food or handling cutlery?”

Score 4: Normal. There is no change compared with before symptom onset, and there has been no change in the type of utensil used (for example chopsticks to knife and fork, or tendency to use a spoon now).

Score 3: Somewhat slow and clumsy, but no help needed. There is some difficulty either cutting food or holding utensils, but the patient is able to do this independently. Use of large handled cutlery or change in utensil used to achieve the task counts as slow or clumsy.

Score 2: Can cut most foods, although slow and clumsy; some help needed. Occasionally assistance is needed from a caregiver, but the patient is independent for the task otherwise.

Score 1: Food must be cut by someone, but can still feed slowly. Assistance is required most of or all the time (more than 50%) for cutting food, but not for feeding. For example, food must be cut but the patient can feed themselves otherwise.

Score 0: Needs to be fed. Assistance is needed for any aspect of the task to be achieved. If someone decides not to cut food or feed themselves but might otherwise be able to, score as 0.
5b. CUTTING FOOD AND HANDLING UTENSILS:

Patients with gastrostomy

If someone has a gastrostomy and it is the primary method of caloric intake, treat as “with gastrostomy”.

Ask “How are you with handling the gastrostomy fastenings and fixtures?”

Score 4: Normal. *Normal means that there is no difficulty at all with any manipulations.*

Score 3: Clumsy, but able to perform all manipulations independently.

Score 2: Some help needed with closures and fasteners.

Score 1: Provides minimal assistance to caregiver.

Score 0: Unable to perform any aspect of task.
6. DRESSING AND HYGIENE

Ask “How are you with dressing or washing?”

Score 4: Normal function. There is no change compared with before symptom onset.

Score 3: Independent. Can complete self-care with effort or decreased efficiency. The person is slower than before but remains independent, and does not use any assistance from either another person or a device such as a button hook.

Score 2: Intermittent assistance or substitute methods. Some help is needed either from a caregiver or by use of devices such as button hooks or self-tying laces, but the patient is otherwise independent. If the patient has changed the clothing they normally wear such as having zipped clothing instead of buttons, score as substitute method.

Score 1: Needs attendant for self-care. All aspects of the task require assistance, but the patient is able to assist the caregiver for much of it.

Score 0: Total dependence. The person is completely unable to carry out any aspect of the task and cannot significantly help the caregiver. If someone decides not to dress or bathe themselves but would otherwise be able to, score 0.
7. TURNING IN BED AND ADJUSTING BED CLOTHES

Ask “Can you turn in bed and adjust the bed clothes?”

Score 4: Normal function.

Score 3: Somewhat slow and clumsy, but no help needed. There is difficulty either with turning in bed or adjusting bed clothes or both.

Score 2: Can turn alone, or adjust sheets, but with great difficulty. There is great difficulty, but the person can perform at least one of the activities independently.

Score 1: Can initiate, but not turn or adjust sheets alone. The process of turning or adjusting bed clothes is begun in some way by the person, but someone else needs to provide the assistance required to complete the task. If one task can be completed independently but not the other, score as 2. If both require assistance to complete, score 1.

Score 0: Helpless. Initiation of turning is impossible.
8. WALKING

Ask “How is your walking?”

Score 4: Normal. *There is no change from walking ability before symptom onset.*

Score 3: Early ambulation difficulties. *There is some difficulty walking, which might include slowing, tripping or imbalance, but no assistance is routinely needed either in the form of help from someone else, or by the use of an ankle-foot orthosis, a walking stick, or frame.*

Score 2: Walks with assistance. *Assistance from a physical aid or a caregiver is needed.*

Score 1: Non-ambulatory functional movement only. *The person can help with transfers by weight bearing.*

Score 0: No purposeful leg movement.
9. CLIMBING STAIRS

Ask “Are you able to climb stairs?”

Only rate ability for walking up stairs, not down.

Score 4: Normal.

Score 3: Slow. *There is some slowing but the person does not rest between steps or feel unsteady.*

Score 2: Mild unsteadiness or fatigue. *The person needs to rest or feels unsteady.*

Score 1: Needs assistance. Use of a handrail or help from a caregiver is required to climb stairs.

Score 0: Cannot do. If someone decides they do not want to climb stairs but would seem otherwise able, score 0.
10. DYSPNOEA

Ask “Do you become breathless?”
Score the patient regardless of the apparent cause of breathlessness.
If someone is using non-invasive ventilation at night or in the day for ALS, score 0.
“Walking” means walking at a comfortable speed on the flat.
Score 4: None.
Score 3: Occurs when walking.
Score 2: Occurs with one or more of the following: eating, bathing, dressing.
Score 1: Occurs at rest: difficulty breathing when either sitting or lying.
Score 0: Significant difficulty: considering using mechanical respiratory support.
11. ORTHOPNOEA

Ask “Can you sleep lying down flat or do you need to be propped up?”

Score based on difficulty regardless of the apparent underlying cause (so for example, needing to sleep sitting up because of excessive saliva scores 1).

Treat a hospital style bed in which the back can be raised independently as if pillows were in place of the raised section.

Score 4: None.

Score 3: Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows. *There is difficulty falling asleep or the person wakes because of breathlessness but they do not use more than two pillows.*

Score 2: Needs extra pillows in order to sleep (more than two). *More than two pillows are needed, or the back is raised up to at least 45 degrees.*

Score 1: Can only sleep sitting up. *The person sleeps sitting up in bed or in a chair.*

Score 0: Unable to sleep without mechanical assistance. *Non-invasive ventilation is used most or all of the night. If NIV is used for an hour or so only, score as if not used.*
12. RESPIRATORY INSUFFICIENCY

Ask “Do you use non-invasive ventilation?”
Regard BiPAP as any form of non-invasive ventilation.
Score 4: None.
Score 3: Intermittent use of BiPAP.
Score 2: Continuous use of BiPAP during the night.
Score 1: Continuous use of BiPAP during day & night.
Score 0: Invasive mechanical ventilation by intubation or tracheostomy.
Practice: using the ALSFRS-R

Following on from this slide are 6 learning activities on more complicated cases for you to test yourself on.

Please try and answer the question before looking at the answer on the next slide.
Question 1: Speech

Please select the correct score for 'speech', based on the interview fragment below.
Interviewer asks : How is your speech?
Patient: “I’m having problems with articulation”.
Interviewer: “Is your speech different to before having any symptoms of ALS?”
Patient: “My speech changed after I started to wear dentures”.

4. Normal speech process
3. Detectable speech disturbance
2. Intelligible with repeating
1. Speech combined with non-vocal communication
0. Loss of useful speech
In case of an obvious external cause like new dentures, the change in speech is not a symptom of ALS. The speech is scored as normal.

The correct answer is:

4. Normal speech process
Question 2: Salivation

Please select the correct score for 'salivation', based on the interview fragment below
Interviewer: “How is your saliva?”
Patient: “Since I started to use medication for it, there is much less saliva than before”.
Interviewer: “Is there still excess of saliva?”
Patient: “Yes but I do not need a tissue for it anymore”.

4. Normal
3. Slight but definite excess of saliva in mouth; may have night time drooling
2. Moderately excessive saliva; may have minimal drooling
1. Marked excess of saliva with some drooling
0. Marked drooling
Answer 2: Salivation

The salivation should be scored as reported, regardless of medication use.

The correct answer is:

3. Slight but definite excess of saliva in mouth; may have night time drooling
Question 3: Handwriting

Please select the correct score for 'handwriting', based on the interview fragment below.

Interviewer: “Are you able to hold a pen?”
Patient: “Yes”
Interviewer: “How is your writing?”
Patient: “I don’t write anymore except if they need a signature”.
Interviewer: “Have you written anything else than your name or signature recently?”
Patient: “No”

4. Normal
3. Slow or sloppy: all words are legible
2. Not all words are legible
1. No words are legible, but can still grip pen
0. Unable to grip pen
Answer 3: Handwriting

The writing should be scored for words except name and signature. If the patient has not written other words except their name or signature recently, score as 1.

The correct answer is:

1. No words are legible, but can still grip pen
Question 4: Patients with gastrostomy

Please select the correct score for 'cutting food and handling utensils: patients with gastrostomy', based on the interview fragment below.

Interviewer: “You have a gastrostomy. Do you eat food through your mouth as well?”
Patient: “No I’m totally fed by the gastrostomy.”
Interviewer: “How are you handling the gastrostomy fastenings and fixtures?”
Patient: “I’m able to handle them myself.”
Interviewer: “Is it clumsy when you handle them?”
Patient: “Yes it goes slow and clumsy. “

4. Normal
3. Clumsy, but able to perform all manipulations independently
2. Some help needed with closures and fasteners
1. Provides minimal assistance to caregiver
0. Unable to perform any aspect of task
Answer 4: Patients with gastrostomy

Cutting food and handling utensils: patients with gastrostomy

In case a patient has a gastrostomy and it is the primary method of caloric intake, treat as ‘with gastrostomy’.

We score the fine motor function by asking about handling the fastenings and fixtures.

The correct answer is:

3. Clumsy, but able to perform all manipulations independently
Question 5: Climbing stairs

Please select the correct score for 'climbing stairs', based on the interview fragment below.

Interviewer: “Are you able to climb stairs?”
Patient: “I don’t have stairs at home”.
Interviewer: “And do you climb stairs elsewhere?”
Patient: “No I’m too scared to climb stairs anymore.”

4. Normal
3. Slow
2. Mild unsteadiness or fatigue
1. Needs assistance
0. Cannot do
Answer 5: Climbing stairs

If someone decides they do not want to climb stairs but would seem otherwise able, score 0.

The correct answer is:

0. Cannot do
Question 6: Orthopnoea

Please select the correct score for 'orthopnoea', based on the interview fragment below.

Interviewer: “Can you sleep lying down flat?”

Patient: “No I use 3 pillows to support my neck. When lying flat, my neck is hurting because of the muscle weakness.”

4. None
3. Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows
2. Needs extra pillows in order to sleep (more than two)
1. Can only sleep sitting up
0. Unable to sleep without mechanical assistance
Answer 6: Orthopnoea

Score as reported, regardless of the apparent underlying cause.

The correct answer is:

2. Needs extra pillows in order to sleep (more than two)
References and other documents:

• ENCALS SOP for ALSFRS-R administration V1.2 10 May 2015
• Revised ALS Functional Rating Scale (ALSFRS-R) MND SMART Training slides
• ALS-Functional-Rating-Scale-Revised-fill-in-form