Revised ALS Functional Rating Scale (ALSFRS-R)

Standard operating procedure

The ALSFRS-R is a scale designed to assess function at home as rated by the patient. The patient should not be prompted in any way, except as described in the SOP, either by the person administering the scale or by a caregiver. 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.

These notes are designed to help clarify some of the ambiguities that can arise in the administration of this scale. The initial question is 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).

ALSFRS-R Questions

1. SPEECH
Ask “How is your speech?” Normal speech means it is exactly the same as before the onset of ALS symptoms. “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. “Intelligible with repeating” means that >25% of the time, repeating is necessary for comprehension. “Speech combined with non-vocal communication” means that writing, use of speech synthesizers or similar methods are needed to supplement speech.

4 Normal speech process
3 Detectable speech disturbance
2 Intelligible with repeating
1 Speech combined with non-vocal communication
0 Loss of useful speech

2. SALIVATION
Ask “How is your saliva?” and score as reported regardless of medication use. Some people have a dry mouth and may report that; if there is no excess saliva, score as normal. “Slight saliva” with or without night time drooling, means that there is an excess, but there is usually no need to mop up the saliva with a tissue. “Moderately excessive saliva” means that a tissue needs to be used, but <25% of the time. “Marked excess of saliva” means that there is likely to be drooling and a tissue is often, but not always used. “Marked drooling” requires a constant use of tissue or handkerchief, or suction.

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

3. SWALLOWING
Ask “How is your swallowing?” Normal means that there is no change from before symptom onset; they should be able to eat any food in typical mouthful sizes or drink liquid without difficulty. “Early eating problems” means that occasionally food will stick, or cause coughing or choking. Food may need to be cut up small, but is not mashed or liquidized. “Dietary consistency changes” means that food needs to be mashed or liquidized, drinks may need thickener, or some foods such as steak, dry biscuits or cornflakes are avoided in favour of yoghurts, casseroles or porridge. “Needs supplemental tube feeding” means that oral intake of food is so difficult that significant weight loss (>10%) has occurred and gastrostomy is required to supplement caloric intake *regardless of whether one is fitted or not*. NPO is exclusively parental or enteral feeding.

4 Normal eating habits
3 Early eating problems – occasional choking
2 Dietary consistency changes
1 Needs supplemental tube feeding
0 NPO

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. “Slow or sloppy, all words are legible” means that using a normal pen there is a change in writing. The person may need to use large pen grips or other writing aids. “Not all words are legible” means that some words cannot be read but others can. Ignore ability to write name or sign legibly. If the patient can only write their name or sign legibly, but other writing is illegible, score as 1. If the patient has not written other words except their name or signature recently and therefore cannot answer the question further, score as 1.

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

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?” Normal means that 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). “Somewhat slow and clumsy, but no help needed” means that there is some difficulty either cutting food or holding utensils, but the patient is able to do this independently. Use of large handled cutlery to achieve the task counts as slow and clumsy. “Can cut most foods although slow and clumsy; some help needed” means that occasionally assistance is needed, but the patient is independent for the task otherwise. “Food must be cut by someone but can still feed slowly” means that assistance is required at least half the time for cutting but not for feeding. For example, if food must be cut but the patient can feed themselves otherwise, score 1. “Needs to be fed” means that assistance is needed for any aspect of the task to be achieved. It someone decides not to cut food or feed themselves but might otherwise be able to, then score 0.

4 Normal
3 Somewhat slow and clumsy, but no help needed
2 Can cut most foods (> 50%), although slow and clumsy; some help needed
1 Food must be cut by someone, but can still feed slowly
0 Needs to be fed

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?” Normal means that there is no difficulty at all with any manipulations.

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

6. DRESSING AND HYGIENE

Ask “How are you with dressing or washing?” Normal means there is no change compared with before symptom onset. “Independent but with effort or decreased efficiency” means 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. “Intermittent assistance or substitute methods” means that 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. “Needs attendant for self-care” means that all aspects of the task require assistance, but the patient is able to assist the caregiver for much of it “Total dependence” means that the patient 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.

4 Normal function
3 Independent; Can complete self-care with effort or decreased efficiency
2 Intermittent assistance or substitute methods
1 Needs attendant for self-care
0 Total dependence

7. TURNING IN BED AND ADJUSTING BED CLOTHES

Ask “Can you turn in bed and adjust the bed clothes?” If there is difficulty with either or both, then rate 3. If there is great difficulty, as long as the patient can perform at least one of the activities independently, rate 2. “Can initiate, but not turn or adjust sheets alone” means that the process of turning 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. “Helpless” means that initiation of turning is impossible.

4 Normal function
3 Somewhat slow and clumsy, but no help needed
2 Can turn alone, or adjust sheets, but with great difficulty
1 Can initiate, but not turn or adjust sheets alone
0 Helpless

8. WALKING

Ask “How is your walking?” Normal means that there is no change from walking ability before symptom onset. “Early ambulation difficulties” means that 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. If assistance from a physical aid or caregiver is needed, score 2. If the patient can help with transfers by weight bearing, score 1.

4 Normal
3 Early ambulation difficulties
2 Walks with assistance
1 Non-ambulatory functional movement only
0 No purposeful leg movement

9. CLIMBING STAIRS

Ask “Are you able to climb stairs?” Only rate ability for walking up stairs, not down. Normal means that there is no change from the situation before symptom onset. Slow means there is some slowing but the patient does not rest between steps of feel unsteady. If they do need to rest of feel unsteady, score 2. “Needs assistance” means use of a handrail or help from a caregiver is required to climb stairs. If someone decides they do not want to climb stairs but would seem otherwise able, score 0.

4 Normal
3 Slow
2 Mild unsteadiness or fatigue
1 Needs assistance
0 Cannot do

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.

4 None
3 Occurs when walking
2 Occurs with one or more of the following: eating, bathing, dressing
1 Occurs at rest: difficulty breathing when either sitting or lying
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. If there is difficulty falling asleep or if the patient wakes because of breathlessness but they do not use more than two pillows, score 3. If more than two pillows are needed, or the back is raised up to at least 45 degrees, score 2. If the patient sleeps sitting up in bed or in a chair, score 1. If non-invasive ventilation is used most or all of the night, score 0. If NIV is used for an hour or so only, score as if not used.

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

12. RESPIRATORY INSUFFICIENCY

Ask “Do you use non-invasive ventilation?” Regard BiPAP as any form of non-invasive ventilation.

4 None
3 Intermittent use of BiPAP
2 Continuous use of BiPAP during the night
1 Continuous use of BiPAP during day & night
0 Invasive mechanical ventilation by intubation or tracheostomy