

# ALSFRS-R Training

ENCALS  
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# Revised ALS Functional Rating Scale (ALSFRS-R) Introduction

- 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).

# Revised ALS Functional Rating Scale (ALSFRS-R) Introduction

- The ALSFRS-R is a scale designed to assess function at home as rated by the patient.
- 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 in the SOP, either by the person administering the scale or by a caregiver.

# Revised ALS Functional Rating Scale (ALSFRS-R) Introduction

- 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.

# 1. SPEECH

- Ask “How is your speech?”

**Score 4; Normal speech process**

Speech is exactly the same as before the onset of ALS 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

# 1. SPEECH

- Ask “How is your speech?”.

**Score 2; Intelligible with repeating**

>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

## 2. SALIVATION

- Ask “How is your saliva?”

**Score 2; Moderately excessive saliva; may have minimal drooling**  
A tissue needs to be used, but <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 liquidized

### 3. SWALLOWING

- Ask “How is your swallowing?”

#### **Score 2; Dietary consistency changes**

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

#### **Score 1; Needs supplemental tube feeding**

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*

#### **Score 0; NPO**

Exclusively parental 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

## 4. HANDWRITING

- Ask “Are you able to hold a pen?”

**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)

## 5a. CUTTING FOOD AND HANDLING UTENSILS: Patients without gastrostomy

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

**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

## 5a. CUTTING FOOD AND HANDLING UTENSILS: Patients without gastrostomy

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

**Score 1;** Food must be cut by someone, but can still feed slowly

Assistance is required most of or all the time (> 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

## 5b. CUTTING FOOD AND HANDLING UTENSILS: Patients with gastrostomy

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

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

## 6. DRESSING AND HYGIENE

- Ask “How are you with dressing or washing?”

### **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

## 6. DRESSING AND HYGIENE

- Ask “How are you with dressing or washing?”

### **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 bedclothes 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

## 7. TURNING IN BED AND ADJUSTING BED CLOTHES

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

**Score 1; Can initiate, but not turn or adjust sheets alone**

The process of turning or adjusting bedclothes 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

## 8. WALKING

- Ask “How is your walking?”

**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

## 9. CLIMBING STAIRS

- Ask “Are you able to climb stairs?”

Only rate ability for walking *up stairs*, not down.

**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.

## 10. DYSPNOEA

- Ask “Do you become breathless?”

Score 4; None

Score 3; Occurs when walking

Score 2; Occurs with one or more of the following: eating, bathing, dressing

# 10. DYSPNOEA

- Ask “Do you become breathless?”

**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.

# 11. ORTHOPNOEA

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

**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

# 11. ORTHOPNOEA

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

**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