ALS Functional Rating Scale (ALSFRS-R)

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Objectives

- Revised ALS Functional Rating Scale (ALSFRS-R) Introduction
- Additional explanation, per category:
  - Bulbar function (*question 1 - 3*)
  - Fine motor function (*question 4 - 6*)
  - Gross motor function (*question 7 - 9*)
  - Respiratory function (*question 10 - 12*)
- Discuss complicated cases.

Revised ALS Functional Rating Scale (ALSFRS-R) Introduction

- The ALS Functional Rating Scale-Revised (ALSFRS-R) is a 12-item functional scale about ability of the ALS patient to perform daily activities. The scale can be divided to 4 subgroups:
- fine motor function,
- gross motor function,
- bulbar function,
- respiratory function

Each item (a specific question about the ability of an ALS patient) is scored from 4 (normal) to 0 (unable).

The ALSFRS-R is a widely used validated instrument for assessment of disease progression in clinical practice and also in clinical trials for ALS as primary or secondary outcome measure.

Revised ALS Functional Rating Scale (ALSFRS-R) Introduction

- ALSFRS-R is simple to administer- in person (clinic), over the phone
- ALSFRS-R can be used for clinical staging (reliably estimated)
- ALSFRS-R assessment gives a quick summary which can be used to identify health care needs of patients (QOL)
- ALSFRS-R score has strong correlation with survival and disease progression
- The ALSFRS-R includes a widespread range of signs and symptoms of ALS

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.
- 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. (use speaker function)

Revised ALS Functional Rating Scale (ALSFRS-R) Introduction

- 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.

Bulbar function *(question 1 - 3)*

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1. SPEECH: Ask "How is your speech?"

4 Normal speech process: *Means it is exactly the same as before the onset of ALS symptoms.*

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.*

2 Intelligible with repeating: *Means that >25% of the time, repeating is necessary for comprehension.*

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

0 Loss of useful speech

Bulbar function *(question 1 - 3)*

2. SALIVATION: Ask "How is your saliva?", score as reported regardless of medication use

4 Normal: *Means if there is no excess saliva.*

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

2 Moderately excessive saliva; may have minimal drooling: *Means that a tissue needs to be used, but <25% of the time.*

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

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

Bulbar function *(question 1 - 3)*

3. SWALLOWING: Ask "How is your swallowing?"

4 Normal eating habits *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.*

3 Early eating problems – occasional choking: *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.*

2 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.*

1 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.*

0 NPO: *is exclusively parental or enteral feeding.*

** Please be aware this differ from the NEALS Standard Operating Procedure.*

Bulbar function *(question 1 - 3)*

Fine motor function *(question 4 - 6)*

Fine motor function *(question 4 - 6)*

4. **HANDWRITING :** Ask “Are you able to hold a pen?” If the answer is “Yes” then ask “How is your writing?” *Only score the dominant hand and only score for use of a standard pen of normal size*

4 Normal: *only score for use of a standard pen of normal size.*

3 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.*

2 Not all words are legible: *means that some words cannot be read but others can. Ignore ability to write name or sign legibly*

1 No words are legible, but can still grip pen*: If the patient can only write their name or sign legibly, but other writing is illegible score as 1.*

0 Unable to grip pen

Fine motor function *(question 4 – 6)*

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?"

4 Normal :*means that there is no change compared with before symptom onset, and there has been no change in the type of utensil used.*

3 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.*

2 Can cut most foods (> 50%), although slow and clumsy; some help needed: *means that occasionally assistance is needed, but the patient is independent for the task otherwise.*

1 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. If food must be cut but the patient can feed themselves otherwise, score 1.*

0 Needs to be fed: *means that 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, then score 0.*

Fine motor function *(question 4 – 6)*

5b.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?"

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

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

Fine motor function *(question 4 – 6)*

6.DRESSING AND HYGIENE : Ask "How are you with dressing or washing?"

4 Normal function: *means there is no change compared with before symptom onset*

3 Independent: Can complete self-care 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.*

2 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 .*

1 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.*

0 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.*

Fine motor function (*question 4 – 6*)

Gross motor function (*question 7 - 9*)

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7. TURNING IN BED AND ADJUSTING BED CLOTHES: Ask "Can you turn in bed and adjust the bed clothes?"

4 Normal function

3 Somewhat slow and clumsy, but no help needed: *If there is difficulty with either or both, then rate 3.*

2 Can turn alone, or adjust sheets, but with great difficulty: *If there is great difficulty, as long as the patient can perform at least one of the activities independently, rate 2.*

1 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.***

0 Helpless: *means that initiation of turning is impossible.*

Gross motor function (*question 7 – 9*)

8. WALKING: Ask "How is your walking?"

4 Normal: *means that there is no change from walking ability before symptom onset.*

3 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.*

2 Walks with assistance: ***If assistance from a physical aid or caregiver is needed, score 2.***

1 Non-ambulatory functional movement only: *if the patient can help with transfers by weight bearing, score 1.*

0 No purposeful leg movement

Gross motor function (*question 7 - 9*)

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

Gross motor function (*question 7 – 9*)

Respiratory function(*question 10 - 12*)

Respiratory function(*question 10 – 12*)

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.*

4 None

3 Occurs when walking: *means walking at a comfortable speed on the flat surface.*

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*

** Please be aware this differ from the NEALS Standard Operating Procedure.*

Respiratory function(*question 10 – 12*)

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.

4 None

3 Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows: *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.*

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

1 Can only sleep sitting up: *If the patient sleeps sitting up in bed or in a chair, score 1.*

0 Unable to sleep without mechanical assistance: *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.*

Respiratory function(*question 10 – 12*)

- **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**

Respiratory function (*question 10 – 12*)

Q&A *(1/4)*

Question 6. Dressing and hygiene: "how are you with dressing or washing?"

Issue: sitting position for most tasks

Q: How to score a patient who has always chosen to sit down to do a task when they could have done it standing prior to ALS (e.g. putting shoes on).

A: General remark, querying is essential in this situation. Firstly, the rater should thoroughly explore whether the patient always has sat down. Secondly, the rater should thoroughly explore the extent of the need for a given substitute method e.g. sitting down to putting on the shoes: can or can not stand up putting on shoes. If the patient is not able to put on shoes in standing position it is scored as 2. A score of 3 means a decreased efficiency i.e. the person is slower than before but remains independent. And a score of 4 means there is no change compared with before symptom onset.

Q&A *(2/4)*

8: Walking: How is your walking?

Issue: use of Wheelchair

Q: How to score a patient who is using a wheelchair prior to ALS onset?

A: General remark, if the patient used a wheelchair prior to ALS onset the patient will be scored as a 2, since physical aid is needed. But query further if the patient is still able to walk a view step, can help with transfer by weight bearing or not able to perform purposeful leg movement.

Q&A *(3/4)*

9. Climbing Stairs: "are you able to climb stairs?"

Issue: use of handrail

Q: How to score a patient who used the handrail prior to disease onset?

A: General remark, querying is essential in this situation. Firstly, the rater should thoroughly explore whether the patient always has used the handrail. Secondly, the rater should thoroughly explore the extent of the need for this substitute method. If the patient is not able to climb the stairs without using the handrail, it should be scored as 1. A score of 4 means there is no change compared with before symptom onset.

Q&A *(4/4)*

12. Respiratory insufficiency: "Do you use non-invasive ventilation?"

Issue: use of CPAP

Q: How to score a patient using CPAP prior to ALS onset due to sleep apnoea?

A:General remark, this patient cannot receive a score of 4 (no use of non-invasive ventilation) but should be scored according to the actual use (hours) of CPAP.

ALSFRS-R Certification

1. Log into TRICALS online training portal and complete interactive training modules
   - http://elearning.elevatehealth.eu
     - Username: email address (lower case)
     - Password: Tricals@2018

2. Attend IM or In-Study Web-Ex training session

3. Complete video examination

4. Obtain certification from TRICALS and please sent it to: smp_orz_3364_cat@worldwide.com

CONCLUSION

- ALSFRS-R is a sensitive questionnaire based measure of changes in physical function decline and disability in ALS patients.
- ALS is a heterogeneous disease : ALSFRS-R demonstrates variable individual patient onset site, spreading pattern and progression
- If the ALSFRS-R scores are incorrect, the data could bias the outcome of randomized clinical trials.
- If ALSFRS-R is conducted in a different way, scores might differ and could not be used as a reliable assessment of function loss in ALS.
- A standard to perform the ALSFRS-R in all trial centres enjoying the TRICALS community.