

Appendix (E)A-1

Self-Administered ALS Functional Rating Scale-Revised

Patient Name: _____ Date: _____

The ALS Functional Rating Scale-Revised is used to assess changes in physical functioning in persons with Amyotrophic Lateral Sclerosis (ALS). This scale is widely used by doctors and health care practitioners in clinical research trials and patient care in the ALS community.

The following questions refer to how you are currently functioning at home. Please read each item carefully and base your answers on your functioning today compared to the time before you had any symptoms of ALS. Please choose the answer that best fits your functional status today. Place an "x" in the box next to your answer.

Compared to the time before you had symptoms of ALS:

1. Have you noticed any changes in your **speech**?

- no change**
- noticeable speech difference**
- speech has changed; **asked often to repeat words or phrases**
- speech has changed; sometimes need the use of **alternative communication methods** (i.e. computer, writing pad, letter board or eye chart)
- unable** to communicate verbally

2. Have you noticed any changes (increases) in the amount of **saliva** in your mouth (regardless of any medication use)?

- no change**
- slight but definite excess of saliva with or without **night time drooling**
- moderate amounts of excessive saliva with or without **minimal day time drooling**
- marked amounts of excessive saliva with **some daytime drooling**
- marked excessive saliva with **marked drooling** requiring a constant tissue or handkerchief

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Compared to the time before you had symptoms of ALS:

3. Have there been any changes in your ability to **swallow**?

- no changes** (all foods and liquids)
- some changes in swallowing or **occasional choking episodes** (including coughing during swallowing)
- unable to eat all consistencies of food and have **modified the consistency of foods** eaten
- use a **feeding tube (PEG) to supplement** what is eaten by mouth
- do not eat anything by mouth** and receive all nutrition through a feeding tube (PEG)

4. Has your **handwriting** changed? Please choose the best answer that describes your handwriting with your **dominant (usual)** hand without a cuff or brace.

- no changes**
- slower and/or sloppier but all the words are **legible**
- not all words are legible**
- able to **hold a pen** but **unable to write**
- unable to hold a pen**

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5. The following question refers to your ability to **cut foods and handle utensils** (feed yourself) compared to before you had symptoms of ALS. If most of your nutrition is through a feeding tube (PEG), skip to **part b** of this question. If you eat most of your meals by mouth answer **part a**.

Compared to the time before you had symptoms of ALS:

a. Cutting food and handling utensils:

- no change**
- somewhat slow and clumsy** (or different than before) but no assistance or adaptive equipment
- sometimes need help** with cutting more difficult foods
- food must be cut by someone else** but can feed slowly without assistance
- need to be fed**

b. Using a feeding tube (PEG)

- use PEG **without assistance or difficulty**.
- use PEG without assistance however may be **slow and /or clumsy**.
- require assistance with closures and fasteners**.
- provide minimal assistance to caregiver**
- unable to perform** any of the manipulations

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Compared to the time before you had symptoms of ALS:

6. Has your ability to **dress and perform self-care activities** (i.e. bathing, teeth brushing, shaving, combing your hair, other hygienic activities) changed?

- no change**
- perform self-care activities without assistance but **with increased effort or decreased efficiency**
- require **intermittent assistance or use different methods** (i.e. sit down to get dressed, fasten buttons with a fastener or your non-dominant hand)
- require daily assistance**
- do not perform self-care activities and **completely dependent** on caregiver

7. Has your ability to **turn in bed and adjust the bed clothes** (i.e.. cover yourself with the sheet or blanket) changed?

- no change**
- can turn in bed and adjust the bed clothes **without assistance** but it is **slower or more clumsy**
- can turn in bed **or** adjust the bedclothes **without assistance but with great difficulty**
- can **initiate** turning in bed or adjusting the bed clothes but **require assistance** to complete the task
- helpless** in bed

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Compared to the time before you had symptoms of ALS:

8. Has your ability to **walk** changed?

- no change**
- walking has changed but do not require any assistance or devices** (i.e. foot brace, cane, walker)
- require assistance** to walk (i.e. cane, walker, foot brace or hand held assistance)
- can move legs or stand up but unable to walk** from room to room
- cannot walk** or move my legs

9. Has your ability to **climb stairs** changed?

- no change**
- slower**
- unsteady** and/or more **fatigued**
- require assistance** (i.e. using the handrail, cane or person)
- cannot climb stairs**

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Compared to the time before you had symptoms of ALS:

10. Do you experience **shortness of breath** or have **difficulty breathing**?

- no change**
- shortness of breath only **with walking**
- shortness of breath with **minimal exertion** (i.e. talking, eating, bathing or dressing)
- shortness of breath **at rest** while either sitting or lying down
- significant shortness of breath** (all of the time) and considering using mechanical ventilation

11. Do you experience shortness of breath or have difficulty breathing while lying down on your back?

- no change**
- occasional shortness of breath while lying on back but **don't routinely use more than two (2) pillows** to sleep
- shortness of breath while lying on back and **require more than two pillows** (or an equivalent) **to sleep**
- can only sleep sitting up** due to shortness of breath
- require the use of respiratory (breathing) support** (BiPAP® or invasive ventilation via tracheostomy) to sleep and **do not sleep without it**

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Compared to the time before you had symptoms of ALS:

12. Do you require **respiratory (breathing) support**?

- no respiratory support**
- intermittent use of BiPAP®**
- continuous use of **BiPAP® at night**
- continuous use of **BiPAP® at night and during the day**
(nearly 24 hours per day)
- mechanical ventilation** by intubation or tracheostomy

Please indicate who completed this survey:

- Patient
- Patient with assistance
- Patient with assistance from caregiver or family member
- Caregiver alone

(BiPAP® is commonly used to describe non-invasive positive pressure ventilation and its use here in no way endorses or promotes a particular product)

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Scoring Sheet (for use by the health care provider)

| | |
|---|--|
| <p>1. SPEECH</p> <p>No change value = 4 Noticeable speech disturbance value = 3 Asked often to repeat words or phrases value = 2 Alternative communication methods value = 1 Unable to communicate verbally value = 0</p> <p style="text-align: right;">Q1. Score =</p> | <p>7. TURNING IN BED AND ADJUSTING BEDCLOTHES</p> <p>No change value = 4 Slower or more clumsy, without assistance value = 3 Can turn alone <u>or</u> adjust bed clothes value = 2 Can initiate but requires assistance value = 1 Helpless in bed value = 0</p> <p style="text-align: right;">Q7. Score =</p> |
| <p>2. SALIVATION</p> <p>No change value = 4 Slight excess saliva, nighttime drooling value = 3 Moderately excessive saliva, minimal drooling value = 2 Marked excess of saliva, some drooling value = 1 Marked drooling, requires constant tissue value = 0</p> <p style="text-align: right;">Q2. Score =</p> | <p>8. WALKING</p> <p>No change value = 4 Change in walking, no assistance or devices value = 3 Requires assistance to walk value = 2 Can move legs or stand up, unable to walk from room to room value = 1 Cannot walk or move legs value = 0</p> <p style="text-align: right;">Q8. Score =</p> |
| <p>3. SWALLOWING</p> <p>No change value = 4 Occasional choking episodes value = 3 Modified the consistency of foods value = 2 Supplemental tube feedings value = 1 NPO (do not eat anything by mouth) value = 0</p> <p style="text-align: right;">Q3. Score =</p> | <p>9. CLIMBING STAIRS</p> <p>No change value = 4 Slower value = 3 Unsteady and/or more fatigued value = 2 Requires assistance value = 1 Cannot climb stairs value = 0</p> <p style="text-align: right;">Q9. Score =</p> |
| <p>4. HANDWRITING</p> <p>No change value = 4 Slow or sloppy, all words legible value = 3 Not all words legible value = 2 Able to hold pen, unable to write value = 1 Unable to hold pen value = 0</p> <p style="text-align: right;">Q4. Score =</p> | <p>10. DYSPNEA</p> <p>No change value = 4 Occurs only with walking value = 3 Occurs with minimal exertion value = 2 Occurs at rest, either sitting or lying value = 1 Significant shortness of breath considering mechanical support value = 0</p> <p style="text-align: right;">Q10. Score =</p> |
| <p>5a. CUTTING FOOD AND HANDLING UTENSILS (patients without gastrostomy)</p> <p>No change value = 4 Somewhat slow and clumsy, needs no help value = 3 Sometimes needs help value = 2 Foods cut by someone else value = 1 Needs to be fed value = 0</p> <p style="text-align: right;">Q5a. Score =</p> | <p>11. ORTHOPNEA</p> <p>No change value = 4 Occasional shortness of breath, does not routinely use more than two pillows value = 3 Require more than 2 pillows to sleep value = 2 Can only sleep sitting up value = 1 Require the use of respiratory support (BiPAP®) to sleep value = 0</p> <p style="text-align: right;">Q11. Score =</p> |
| <p>5b. CUTTING FOOD AND HANDLING UTENSILS (patients with gastrostomy)</p> <p>Uses PEG without assistance or difficulty value = 4 Somewhat slow and clumsy, needs no help value = 3 Requires assistance with closures and fasteners value = 2 Provides minimal assistance to caregiver value = 1 Unable to perform any manipulations value = 0</p> <p style="text-align: right;">Q5b. Score =</p> | <p>12. RESPIRATORY INSUFFICIENCY</p> <p>No respiratory support value = 4 Intermittent use of BiPAP® value = 3 Continuous use of BiPAP® at night value = 2 Continuous use of BiPAP day and night value = 1 Invasive mechanical ventilation value = 0</p> <p style="text-align: right;">Q12. Score =</p> |
| <p>6. DRESSING AND HYGIENE</p> <p>No change value = 4 Performs without assistance with increased effort or decreased efficiency value = 3 Intermittent assistance or different methods value = 2 Requires daily assistance value = 1 Completely dependent value = 0</p> <p style="text-align: right;">Q6. Score =</p> | <p style="text-align: right;">Total Score = _____ / 48</p> |