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ORIGINAL ARTICLE

Timing of Communication Device Introduction Defined by ALSFRS-R Score in Patients with Amyotrophic Lateral Sclerosis

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Objective: Augmentative and alternative communication (AAC) devices are crucial for amyotrophic lateral sclerosis (ALS) patients because disease progression impairs verbal speech. Although the introduction of AAC devices must be appropriately timed, no guidelines currently exist. In this study, we examined the usefulness of the ALS functional rating scale-revised (ALSFRS-R) for predicting the timing of device introduction. **Methods:** This study was a retrospective cross-sectional study with consecutive sampling of patients diagnosed with ALS who underwent rehabilitation at Kitasato University East Hospital between 2011 and 2018. Patients were introduced to AAC devices (writing, communication boards, switch control, and/or eye control) and underwent assessment at three timepoints: the start of rehabilitation, as each communication device was introduced, and at the end of rehabilitation. ALSFRS-R multiple comparisons were analyzed using the Kruskal-Wallis test and, as a post-test, the Steel-Dwass test was used. Receiver operating characteristic (ROC) curves and areas under the ROC curves (AUCs) based on ALSFRS-R total and sub-item scores were used to calculate cut-off values for when transitioning to a new type of device is necessary. **Results:** In this study, 216 patients underwent rehabilitation, and 92 met the inclusion criteria. The total ALSFRS-R scores significantly differed among the four devices, except for those between communication boards and switch control devices. The bulbar and respiratory sub-scores did not significantly differ between devices. For each device type, total or sub-item scores yielded an AUC of 0.8 or more. **Conclusions:** Our findings suggest that the ALSFRS-R is a useful assessment for timing the introduction of communication devices, and its utilization could help therapists, caregivers, and families to provide AAC for patients with ALS.

Key Words: augmentative and alternative communication (AAC); communication tools; occupational therapy; rehabilitation

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is characterized by the progressive degeneration of upper and lower motor neurons, leading to limb paralysis, dysphagia, dysarthria, and respiratory failure. Patients can become vulnerable when disease progression results in the loss of speech and motor functions, potentially forcing them to abdicate decision making.[1)] Although patient autonomy could be preserved by introducing communication devices such as writing devices, communication boards, eye control devices, and switch control devices, their operation requires adequate motor functions, and these will inevitably decline over time. Writing devices require upper motor function to hold a pencil and control the upper limb, communication boards require upper motor function or eye movements and blinks, eye control devices require eye movement, and switch devices have various types of switches, including gripping, pushing, and

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touching. Switch devices can be used as long as the ability to activate the switch remains, such as with eyelid, tongue, finger, or toe movement.

The 5-year survival rate for patients with ALS is 8.8% for bulbar onset, 20% for limb onset, 52% for flail arm syndrome, and 64% for flail leg syndrome.[2] Because survival is so variable, providing a clear diagnosis and prognosis and discussing augmentative and alternative communication (AAC) should begin as early as possible[3] to better manage ALS patients and reduce caregiver burden.[4] Accordingly, therapists must judge the appropriate timing for the introduction of a communication device; however, no professional guidelines clearly state when the devices should be introduced.

Function and activities of daily living assessments are made in ALS patients using the Modified Norris Scale,[5] the ALS Assessment Questionnaire,[6] and the ALS Functional Rating Scale-Revised (ALSFRS-R).[7] The ALSFRS-R rates 12 items across four functional domains: bulbar, fine motor, gross motor, and respiratory. Because the ALSFRS-R is self-reported or observationally scored by therapists, it is easier to use in clinical settings than the Modified Norris Scale; furthermore, the ALSFRS-R score is widely used as a primary outcome measure in ALS clinical practice and clinical trials[8] because it provides a superior assessment of disease severity and disease progression. Moreover, the ALSFRS-R can predict survival time by comparing the ratio of scores between the first emergence of symptoms and the first examination, during the entire disease course, or within 100 days.[9]

Despite the validity and utility of the ALSFRS-R, no studies have examined its ability to inform the introduction of communication devices. The purpose of this study was to examine the usefulness of the ALSFRS-R to predict the appropriate timing of communication device introduction.

METHODS

Design and Participants

In this retrospective, cross-sectional study, we used consecutive sampling to collect data from April 1, 2011, to March 31, 2018. Patient inclusion criteria were diagnosis with ALS and the receipt of rehabilitation at Kitasato University East Hospital. ALS patients were diagnosed as "Probable" or above on the Awaji diagnostic criteria.[10] Exclusion criteria included the absence of an ALSFRS-R score, the diagnosis of dementia, or a score of 20 or less on Hasegawa's Dementia Scale-Revised.[11] We collected data retrospectively on clinical demographics (age, gender), ALS disease type (upper limb onset, lower limb onset, and bulbar onset), artificial ventilator usage (noninvasive positive pressure ventilation and tracheotomy positive pressure ventilation), and ALSFRS-R scores. All patients were prescribed rehabilitation by a neurologist and underwent standard occupational therapy, speech therapy, or physiotherapy while in hospital or as an outpatient. The frequency of rehabilitation during hospitalization was daily, whereas for outpatients it was at least once a month. Patients underwent rehabilitation with one or more therapists throughout the progression of disease, and the length of rehabilitation varied according to the prognosis.

Participants were classified as either receiving introduction to communication devices or not receiving such an introduction. The operational definition of "introduction to communication devices" included both the suggestion and provision of devices to the participants. To evaluate the introduced timing of each device in this study, we considered participants who used different devices at different times to be separate participants.

The sample size of this study was based on the desired statistical power (80%) to detect an area of 0.8 under the receiver operating characteristic (ROC) curve (AUC) of the ALSFRS-R, with a two-sided α of 5%. A sample size totaling 20 (introduced: 10 subjects, not introduced: 10 subjects) was derived by insertion of 1-power (0.80), α (0.05), and AUC (0.80) values in the Goksuluk web tool for ROC curve analysis.[12] We therefore planned to collect more than 10 patients' data for each group, i.e., at least 10 with and 10 without communication device introduction. This study was approved by the Clinical Research Review Board of The Kitasato Institute (B18-065), and was performed in accordance with the Declaration of Helsinki.

Communication Devices

The communication devices used by patients in this study included writing devices, communication boards, switch control devices, and eye control devices (**Fig. 1**). The nine occupational therapists and five speech therapists performing device introduction are retrained on communication devices annually, and the introduction of communication devices was considered at the inter-professional meeting for ALS patients.

Assessment Tools

ALSFRS-R was used to examine its utility for evaluating the timing of communication device introduction. Each of the 12 items that make up the ALSFRS-R are rated on a five-item scale that correspond to scores from 0 to 4. The

Writing devices	Communication board	Switch control devices	Eye control devices
・Boogie board ・KakiPon®	・Message board ・Kana Board	・Talking aid ・Den-no-shin®	・miyasuku EyeCon SW® ・My Tobii®

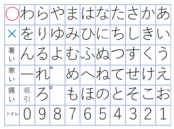

Fig. 1. Examples of communication devices.

total scores therefore range from 0–48, and a lower score indicates a worse condition, e.g., speech: 4 (normal speech processes), 3 (detectable speech disturbance), 2 (intelligible with repeating), 1 (speech combined with non-vocal communication), and 0 (loss of useful speech). We divided the 12 items into 4 groups: the bulbar sub-score included items 1 to 3 (speech, salivation, and swallowing), the upper motor sub-score included items 4 to 6 (handwriting, cutting food and handling utensils, and dressing and hygiene), the gross motor sub-score included items 7 to 9 (turning in bed and adjusting bed clothes, walking, and climbing stairs), and the respiratory sub-score included items 10 to 12 (dyspnea, orthopnea, and respiratory insufficiency). The reliability and validity of the ALSFRS-R have been previously established.[7)]

To find suitable cut-off values between patients who were introduced to a communication device and those who were not, ALSFRS-R scores were assessed at the start of rehabilitation, as each communication device was introduced, and at the end of rehabilitation. Patients who were not introduced to communication devices were allocated the ALSFRS-R score at the end of rehabilitation. In our analyses, to evaluate the timing of introduction of each device, we refer to device introduction as "introduced," and the absence of device introduction as "not introduced." Patients who used different devices at different times were analyzed as if they were separate patients for each device. For example, for writing devices, ALSFRS-R scores were categorized as "introduced" for patients who were introduced to a writing device, and "not introduced" for patients who were introduced to a communication board, switch control device, or eye control device. Patients who were not introduced to a device before the end of rehabilitation were classified as "not introduced". In this study, we determined the cut-off values between devices to know when to shift from one device to another.

Statistical Analysis

Median ALSFRS-R scores were calculated for each device and disease type, and multiple comparisons were analyzed using the Kruskal-Wallis test with the Steel-Dwass test as a post-test. Additionally, to find cut-off values for ALSFRS-R total scores and sub-scores (bulbar, upper motor, gross motor, and respiratory), we examined the ROC curves of each device. The AUC helped us to determine the ideal time for device transition by measuring the quality of the model's predictions. Generally, an AUC of 0.7–0.8 is acceptable, 0.8– 0.9 is excellent, and more than 0.9 is outstanding.[13,14)] Moreover, sensitivity and specificity were used to examine the accuracy of the ROC curve to define the cut-off values for device introduction. All statistical analyses were conducted using JMP software (SAS Institute, version 14.0.0, Marlow, UK), and statistical significance was set at $P < 0.05$.

RESULTS

Characteristics of the Participants and Communication Devices

In this study, 216 patients underwent rehabilitation at Kitasato University East Hospital between April 1, 2011, and March 31, 2018. Ninety-two patients were included for data analysis (**Fig. 2**, **Table 1**). We observed no significant differences among the type of ALS and the timing of communication device suggestion or provision ($P>0.05$; **Table 1**).

Among the 92 patients included in the current study, a total of 77 devices were introduced (14 writing devices, 27 communication boards, 24 switch control devices, and 12 eye control devices). The total ALSFRS-R score at device introduction significantly differed across devices, except between communication boards and switch control devices (**Table 2**). Upper motor sub-scores were highest among patients using writing devices (8.0) but, as this sub-score declined, patients shifted toward eye control devices. However, bulbar and

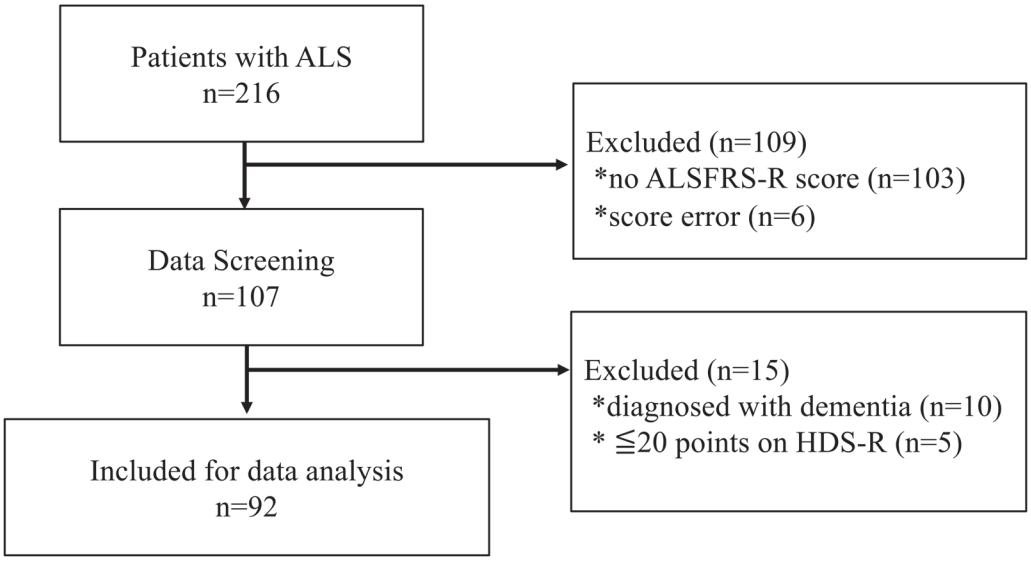

Fig. 2. Flow of patients through the study. HDS-R: Hasegawa's Dementia Scale-Revised.

Table 1. Demographic data for 92 ALS patients (n ± range)

	Overall	Writing devices		Communication boards		Switch control devices		Eye control devices	
	(n=92)	Introduced (n=14)	Not introduced (n=70)	Introduced (n=27)	Not introduced (n=54)	Introduced (n=24)	Not introduced (n=47)	Introduced (n=12)	Not introduced (n=46)
Age at first rehabilitation (years)	67.43 (±9.81)	63.57 (±7.80)	68.79 (±9.64)	65.81 (±9.25)	68.42 (±9.79)	63.59 (±9.57)	69.23 (±9.24)	64.91 (±8.82)	69.67 (±8.82)
Sex									
Male	53	8	40	17	27	12	25	10	25
Female	39	6	30	10	27	12	22	2	21
Type									
Upper limb onset	39	2	34	8	28	11	22	5	21
Lower limb onset	24	3	19	10	14	7	12	4	12
Bulbar onset	27	9	15	9	10	6	11	3	11
Unclear[a]	2	0	2	0	2	0	2	0	2
Artificial ventilator									
None	41	9	30	9	23	9	22	3	22
NPPV	46	4	38	17	29	15	23	7	23
TPPV	5	1	2	1	2	0	2	2	1

[a] Patients were diagnosed at other hospitals and data were not listed in the available charts. NPPV: noninvasive positive pressure ventilation, TPPV: tracheotomy positive pressure ventilation.

respiratory sub-scores did not significantly differ between devices.

Evaluation of ALSFRS-R for Informing Device Introduction

We utilized the area under the ROC curve to determine cut-off values of ALSFRS-R scores that could predict the timing of communication device introduction (**Fig. 3**, **Table 3**). ALSFRS-R score cut-off values worked well for all devices, with each device having a total score or sub-item with an AUC of 0.80 or more. The cut-off values of device introduction timing determined by ROC were as follows: writing

Table 2. ALSFRS-R total and sub-scores [median (inter-quartile range)] at the introduction of communication devices

	Writing devices	Communication boards	Switch control devices	Eye control devices
Total [a,c,f]	32.0 (25.3–33.0)	19.0 (15.0–27.5)	24.0 (19.5–30.5)	13.0 (8.0–16.3)
Bulbar sub-score	6.0 (5.0–7.8)	6.0 (5.0–7.0)	7.0 (5.6–10.0)	5.0 (4.0–6.3)
Upper motor sub-score [b,c,d,e]	8.0 (6.0–9.0)	6.0 (5.0–7.0)	3.5 (0.0–5.0)	0.0 (0.0–0.0)
Gross motor sub-score [a,c]	7.0 (4.0–8.8)	2.0 (0.0–5.0)	3.0 (1.0–7.3)	0.0 (0.0–1.0)
Respiratory sub-score	11.0 (8.3–11.8)	10.0 (7.0–12.0)	11.0 (8.8–12.0)	7.0 (4.0–9.3)

[a] Writing devices vs. communication boards ($P<0.05$), [b] writing devices vs. switch control devices ($P<0.001$), [c] writing devices vs. eye control devices ($P<0.01$), [d] communication boards vs. switch control devices ($P<0.01$), [e] communication boards vs. eye control devices ($P<0.001$), [f] switch control devices vs. eye control devices ($P<0.01$).

devices, upper motor score of 6.0; communication boards, bulbar score of 9.0; switch control devices, respiratory score of 8.0; and eye control devices, total score of 20, bulbar score of 7.0, and gross motor score of 2.0.

DISCUSSION

The purpose of this study was to investigate the usefulness of the ALSFRS-R score, a standard assessment in ALS, to predict the timing of communication device introduction.

In this study, the total ALSFRS-R score at the time of device introduction was significantly different for each device, except between communication boards and switch control devices. This absence of a difference is a result of mixed use of, or transition between, communication boards and switch control devices. This mixed use occurs because communication boards are inexpensive, low tech, and easy to make; consequently, patients can obtain a board during a hospital visit, whereas for switch control devices, patients need to wait months to obtain the device. Consequently, many patients use communication boards while they are waiting for their switch control devices to arrive. Overall, our findings suggested that equipment was introduced according to the remaining motor function as symptoms progressed.

AUC analyses indicated that our ALSFRS-R score cut-off values worked well for all communication devices, with the AUC exceeding 0.80 for each device type. For writing devices, the AUC for the ALSFRS-R upper motor sub-score was 0.80, and the cut-off value was 6.0; because the use of writing devices depends on upper limb function, its introduction was timed to complement the writing ability of ALS patients. The bulbar sub-score AUC for communication board devices was 0.85, and the cut-off value was 9.0; these devices are introduced when swallowing function declines as bulbar paralytic symptoms progress. For switch control devices, the respiratory sub-score AUC was 0.86, and the cut-off value was 8.0. Switch control devices are considered when artificial ventilators become necessary, as reflected by reduced ALSFRS-R respiratory sub-scores. Furthermore, for eye control devices, the AUCs of the total score and the bulbar and gross motor sub-scores were greater than 0.80, with a cut-off value of 2.0 for the gross motor sub-score. This suggests that eye control devices are introduced when gross motor functions are almost completely lost, rendering patients unable to use writing or switch control devices.

For device and sub-score combinations with AUCs greater than 0.80, the sensitivity of tests with the sub-score cut-off values we determined exceeded 80%, except for the respiratory sub-scores for switch control devices. These findings demonstrated the usefulness of ALSFRS-R score cut-off values for informing the timing of communication device introduction. For switch control devices, the sensitivity of the chosen respiratory ALSFRS-R sub-score was low (79.2%); therefore, other measures of respiratory function, such as the sniff nasal inspiratory pressure, should be examined as potential determinants of the timing of device introduction. Moreover, test specificities for some devices were around 60%. This was because successful AAC outcomes rely on various factors, including physical status, cognitive status, language ability, primary user environment, and user motivation,[3] and early introduction may enable patients to better learn to use the equipment. Across devices, errors in the timing of introduction may have occurred due to therapists' evaluation experience, but these events were likely infrequent, given the overall high AUC values for each device.

The validity and reliability of the ALSFRS-R and its relationship with the predictors of ALS progression have been well studied; however, its ability to inform the introduction of communication devices has not been examined. Because this study was retrospective and cross-sectional and the num-

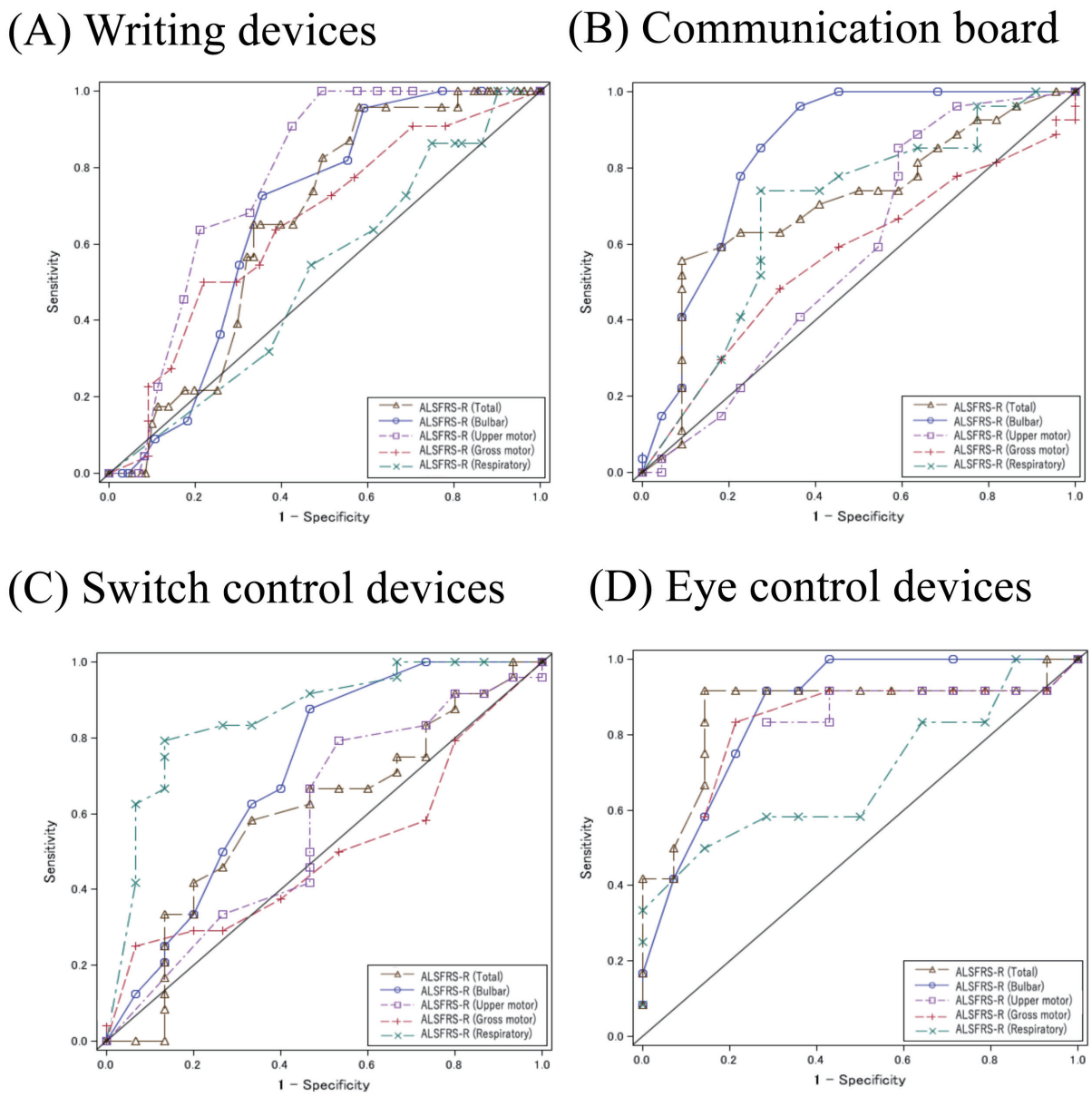

Fig. 3. Receiver operating characteristic (ROC) curve analysis of ALSFRS-R scores for each device. (A) Writing devices, (B) communication boards, (C) switch control devices, (D) eye control devices. The area under the ROC curve for each device conveys its accuracy in differentiating sensitivity and specificity.

ber of devices introduced was small, our test was unable to inform the timing of introduction of switch control devices, likely due to the wide variety of switch control devices used. Additionally, because of the small sample size, we could not examine or exclude the configuration of sub-scores. We believe that additional prospective studies with a larger number of participants will result in more reliable cut-off values to inform device introduction.

It is recommended that the ALSFRS-R score be assessed once a month after the baseline assessment and then used with reference to the cut-off values obtained in this study along with clinical symptoms to help decide the timing of communication device introduction.

The ALSFRS-R is a simple evaluation scale that can be administered by therapists, caregivers, or even families. Our findings suggest that using ALSFRS-R scores could help therapists, caregivers, and families to time the introduction of AAC.

Table 3. Predicting the timing of communication device introduction using ALSFRS-R

Device	ALSFRS-R	AUC	Cut-off value	Sensitivity (%)	Specificity (%)
Writing devices	Total	0.65	30.0	64.3	65.8
	Bulbar sub-score	0.75	9.0	92.9	55.3
	Upper motor sub-score	0.80	6.0	92.9	60.5
	Gross motor sub-score	0.70	8.0	50.0	78.0
	Respiratory sub-score	0.50	10.0	57.1	52.6
Communication boards	Total	0.71	20.0	55.6	90.9
	Bulbar sub-score	0.85	9.0	96.3	63.6
	Upper motor sub-score	0.58	4.0	85.2	40.9
	Gross motor sub-score	0.57	1.0	48.1	66.7
	Respiratory sub-score	0.70	8.0	74.1	72.7
Switch control devices	Total	0.60	24.0	58.3	66.7
	Bulbar sub-score	0.71	10.0	87.5	53.3
	Upper motor sub-score	0.58	5.0	79.2	46.7
	Gross motor sub-score	0.47	8.0	25.0	92.9
	Respiratory sub-score	0.86	8.0	79.2	86.7
Eye control devices	Total	0.86	20.0	91.7	85.7
	Bulbar sub-score	0.87	7.0	91.7	71.4
	Upper motor sub-score	0.76	0.0	83.3	71.4
	Gross motor sub-score	0.81	2.0	83.3	78.6
	Respiratory sub-score	0.69	8.0	50.0	85.7

CONCLUSION

Because of the progressive nature of ALS, assistive communication devices must be introduced with appropriate timing. Therefore, in this study, we examined the usefulness of the ALSFRS-R, a widely used primary outcome measure, as a predictor of device introduction. The ALSFRS-R total score was significantly different across each of the four devices, except for those between communication boards and switch control devices, and all devices had an AUC for the ALSFRS-R total or a sub-item score of 0.8 or greater. Our findings suggest that the ALSFRS-R is a useful assessment for timing communication device introduction that could help therapists, caregivers, and families to provide communication assistance to patients with ALS.

CONFLICTS OF INTEREST

The authors have no conflicts of interest directly relevant to the content of this article.

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