Quality of life and motor impairment in ALS: Italian validation of ALSAQ

A. Palmieri*, G. Sorarù*, L. Lombardi†, C. D'Ascenzo*, L. Baggio*, M. Ermani*, E. Pegoraro* and C. Angelini*

**Department of Neurosciences, University of Padova, Padova, Italy*
†*Department of Cognitive Sciences and Education, University of Trento, Trento, Italy*

Objectives: To evaluate the validity and reliability of the amyotrophic lateral sclerosis assessment questionnaire (ALSAQ) in an Italian cohort of ALS patients and to further characterize the relationship between motor impairment and quality of life (QoL) in ALS.

Methods: Seventy-six patients completed the Italian version of ALSAQ-40 and ALSAQ-5. To verify test–retest reliability, 30 patients were revaluated after 3 months. The medical outcome study short form 36 (MOS SF-36) questionnaire and revised ALS functional rating scale (ALSFR-R) scale were used to assess Italian ALSAQ-40 construct validity. The limb muscles' Medical Research Council (MRC) score and forced vital capacity (FVC) were used to measure the degree of motor impairment.

Results: The Italian ALSAQ-40 showed a very good internal consistency (all subscales Cronbach's alpha>0·86) and a good construct validity as shown by the patterns of correlation between the subscales and SF-36 (resp. ALSFRS-R) scores. ALSAQ-5 showed a positive correlation with the corresponding ALS patient total score and subscale scores of the ALSAQ-40 (Spearman's correlation coefficient>0·73). The emotional functioning subscale did not correlate with any motor impairment measures.

Discussion: Italian ALSAQ-40 and ALSAQ-5 psychometric properties are reliable and similar to those showed by the original English version. We observed emotional aspects to be distinct from physical involvement.

Keywords: ALS, ALSAQ-40, ALSAQ-5, emotional functioning, motor impairment, quality of life

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive disease involving the motor neurons. It has an incidence of ~1·9/100,000 and a prevalence of ~6/100,000[1]. The clinical picture of ALS is characterized by increasing weakness of limbs, trunk, ventilatory and bulbar muscles which usually leads to death after 3–5 years for respiratory failure. As no effective treatment is so far available for ALS, most medical interventions aim to improve the quality of life (QoL) of patients[2]. The QoL in ALS is related to patient psychological profile[3,4], environmental and social condition[5], spiritual aspiration[6] and coping style[7]. The association of life quality with muscle strength and pulmonary function is controversial.

To assess QoL in ALS, the following scales are mostly used: a 36-item medical outcome study short form health survey (MOS SF-36)[8,9], the sickness impact profile/ALS scale (SIP/ALS)[10], the schedule for the evaluation of individual quality of life-direct weighting (SEIQoL-DW)[11], the McGill quality of life questionnaire (MQOL)[12], the amyotrophic lateral sclerosis assessment questionnaire-40-item scale and its shortened 5-item version (ALSAQ-40 and ALSAQ-5 respectively)[13,14]. Unlike the other scales, ALSAQ-40 has been specifically designed for the use in ALS and other motor neuron diseases. It was developed on the basis of in-depth interviews with patients rather than relying on the literature of clinical scales in this field. The ALSAQ-40 reliability of the measures in terms of internal reliability, construct and content validity is high[15,16] and appears sensitive to changes that have an impact on the overall health status of patients. It therefore provides a meaningful and interpretable picture of disease impact on the subjective functioning and well-being of patients on areas that are of concern to them[15]. This scale has been already validated in Japanese[17] and Dutch language[18].

Our purpose is to validate an Italian version of ALSAQ-40 and ALSAQ-5 in a large cohort of Italian ALS patients and to further characterize ALSAQ-40 relation with muscle strength, motor disability and respiratory function.

Patients and methods

Patients

We considered patients affected with clinically definite or clinically probable ALS[19] and regularly

Correspondence and reprint requests to: Arianna Palmieri, PhD, Department of Neurosciences, University of Padova, via Giustiniani 5, 35128 Padova, Italy. [arianna.palmieri@unipd.it]

DOI 10.1179/174313209X385734

followed at the Neuromuscular Clinic of the University of Padova. Exclusion criteria were closeness to death and a level of cognitive impairment not allowing patients to understand the questions.

An informed consent was obtained from each patient.

Italian ALSAQ-40 and ALSAQ-5

Original amyotrophic lateral sclerosis questionnaire contains 40 questions that measure five areas of health: physical mobility (ten items), activities of daily living and independence (ten items), eating and drinking (three items), communication (seven items) and emotional functioning (ten items). The questions refer to the patient's condition during the past 2 weeks and the answers are given on a five-point Likert scale. The ALSAQ-40 indicates the amount of ill health in each domain assessed using a summary score from 0 (the best health status) to 100 (the worss health status). The ALSAQ-5 contains five questions, one from each of the five dimensions of the ALSAQ 40. The score of the five questions of ALSAQ-5 is recoded and ranges from 0 to 100, with 0 representing the best possible health status[13,15,20]. Original questionnaires were translated to Italian by two native Italian speakers fluent in English who have not seen the scales before. The Italian version was back-translated to English by two native English speakers fluent in Italian, who also have not seen the scales before. Since there was no significant difference between the original and back-translated version of the scales, Italian version of scales was accepted. Questionnaire data were collected in a face-to-face setting at the end of the medical assessment. If the patient could write, (s)he filled out the questions of the measure. In the case of writing impairment, the interviewer (psychologist) noted patient's answer to questionnaires. The interviewer noted also any further clarification about questionnaires items requested by the patient. Thirty patients underwent ALSAQ-40 retest by 3 months to analyse the validity and reliability of the Italian version of the scales.

Comparative instruments

To test Italian ALSAQ-40 validity, we used the following measures:

- The amyotrophic lateral sclerosis functional rating scale revised (ALSFRS-R)[21]. This is a 12-item scale, in which the patient's functioning for each item is rated on a scale from 0 (unable to attempt the task) to 4 (normal function). This scale includes evaluation of swallowing, speech and respiratory function, and both strength and function of the upper and lower extremities musculature. A score of 48 points is normal whereas 0 points indicates maximal dysfunction.
- The 36 item short form health survey questionnaire (SF-36)[22]. At present, this is the most widely used measure of general health status. It contains 36 items across eight multi-item scales: physical functioning (PF), role limitations due physical health problems (RP), role limitations due to emotional problems (RE), social functioning (SF), bodily pain (BP), vitality (VT), mental health (MH) and general health perceptions (GH). All raw scales are linearly converted to a 0–100 scale, with higher values indicating favorable health status.
- Medical Research Council (MRC) scale and forced vital capacity (FVC). To evaluate patients' motor disability, we used the measure of muscle strength by the limb muscles: score from 0 (the absence of movement) to 5 (full strength). Muscles evaluated were: deltoids, triceps and biceps brachii, finger extensor, thumb adductor, thigh flexor, knee extensor, ankle extensor and flexor. All muscles were tested bilaterally. We considered the total MRC score (megascore) of both upper and lower limbs which respectively ranged from 0 to 60 and from 0 to 40. FVC was assessed by a standard manual spirometer (CYTEC 60) and expressed as a percentage of the expected value.

Statistical analysis

All data were analysed using the psychometric package as provided in the R language and environment for statistical computing[23]. Usual baseline data and ALS characteristics (including age, gender, type, marital status, time since onset and onset of disease) were evaluated.

The internal reliability of the translated ALSAQ-40 scales and the test–retest reliability were assessed by Cronbach's alpha, as shown in *Table 1*. Non-parametric Spearman's correlation coefficient was used to analyse shortened version, ALSAQ-5, and it was hypothesized that scales with similar content would correlate strongly (0·73 or higher). For the item to scale correlation, corrected for overlap, a correlation coefficient of 0·40 was considered as the required minimum. Construct validity is demonstrated by comparison of the novel rating instruments to other established ones (MOS SF-36, ALSFRS-R) and to functional or instrumental measures (MS upper and lower, FVC) (*Table 2*).

Separate analysis was conducted for the ALSAQ-40 and for the reduced version of the questionnaire (ALSAQ-5).

Moreover, we assessed face validity on the percentage of motor and cognitive help asked.

Table 1 Internal reliability (Cronbach's alpha) of scales of the ALSAQ-40 I in the baseline (time 1) survey and in the retest (time 2)

ALSAQ 40 I scales	Time 1	Time 2
Physical mobility	0·938	0·960
ADL/independence	0·962	0·966
Eating and drinking	0·962	0·965
Communication	0·971	0·965
Emotional functioning	0·886	0·867

Results

Seventy-six patients completed ALSAQ-40 over a period of 20 months. There were no missing data. All patients were interviewed and assessed in hospital; the characteristics of total cohort of patients and their baseline measure are shown in *Tables 3* and *4*. They were similar to expectation and previous report in gender ratio, disease onset and disease type[24].

Internal reliability

The internal reliability involves examining the extent to which a number of items addressing the same concept actually are doing so. Cronbach's alpha values were greater than 0·88 for all dimensions of the Italian ALSAQ-40. Corrected items to subscale correlations were all above 0·60 and significant ($p<0·001$).

Test–retest reliability

Test–retest reliability involves the administration of the measure on two separate occasions to the same population. In our patients, we separate administrations of questionnaires by 3 month follow-up. Internal reliability of scales of the Italian ALSAQ-40 in the

Table 2 Construct validity: subscales of ALSAQ-40 I, ALS-FRS and SF-36

ALSAQ 40	Physical mobility	ADL/independence	Eating and drinking	Communication	Emotional functioning
ALS-FRS subscales					
Speech	0·042	−0·155	−0·718**	−0·795**	−0·234*
Salivation	0·079	−0·115	−0·653**	−0·636**	−0·087
Swallowing	0·033	−0·184	−0·814**	−0·761**	−0·212
Writing	−0·441**	−0·710**	−0·174	−0·151	0·014
Feeding	−0·428**	−0·808**	−0·278*	−0·204	−0·143
Dressing	−0·704**	−0·736**	−0·014	−0·035	−0·118
Turning	−0·739**	−0·707**	−0·085	−0·041	−0·213
Walking	−0·817**	−0·399**	0·041	0·060	−0·223
Climbing	−0·817**	−0·343**	0,077	−0·051	−0·279*
Dyspnea	−0·085	0·007	−0·234*	−0·305**	−0·129
Orthopnea	−0·231*	−0·169	−0·331**	−0·403**	−0·248*
Respiratory insufficiency	−0·261*	−0·142	−0·308**	−0·379**	−0·158
SF-36 subscales					
PF	−0·745**	−0·497**	0,118	0,18	−0·173
RP	−0·160	0·066	−0·156	−0·260	−0·478**
BD	−0·531**	−0·190	0·436**	0·097	0·067
GH	−0·351	−0·196	−0·231	−0·036	−0·345
VT	−0·592**	−0·681**	−0·285	−0·147	−0·582**
SF	−0·518**	−0·483**	−0·004	−0·092	−0·295
RE	−0·046	0·14	0,107	−0·182	−0·507**
MH	−0·008	−0·004	0·062	−0·188	−0·561**
Functional measurement					
FVC	−0·399**	−0·337*	−0·327*	−0·393**	−0·125
MS-sup	−0·397**	−0·709**	−0·139	−0·167	−0·143
MS-inf	−0·651**	−0·332**	0·098	0·110	−0·071

**$p<0·01$, *$p<0·05$.
Spearman correlation coefficient: all correlations are negative as high ALSAQ 40 scores correspond with low ALS-FRS and SF-36 scores.
PF=physical functioning; RP=role limitations due to a physical health problems; RE=role limitations due to an emotional problems; SF=social functioning; BP=body pain; VT=vitality; MH=mental health.

Table 3 Clinical features of 76 ALS patients

Age (years)	Type		Gender		Marital status		Diagnosis		Onset	
76 (mean: 61·39 ± 11·6)	S	F[a]	M	F[b]	Married	Single widow divorced	<2 years	>2 years	B	L
	73	16	39	37	66	10	45	31	15	61

S=sporadic. F[a]=familiar type; M=male; F[b]=female; B=bulbar, L=limb onset.

Table 4 Baseline measures and comparative data

Measurement	No. of patients (%)	Mean	SD
ALSFRS-R	74 (97·4)	33·9	12·1
FVC%	50 (65·8)	79·2	21·8
MS upper	74 (97·4)	47·1	15
MS lower	74 (97·4)	28·5	11·8
SF-36	21 (27·6)		
ALSAQ-40 retest	30 (39·5)		
Questionnaire completed with motor help from someone else	25 (32·9)		
Questionnaire completed with cognitive help from someone else	43 (56·6)		
Questionnaire completed without any kind of aid	13 (17·1)		

SD=standard deviation; FVC: forced vital capacity; ALSFRS-R: amyotrophic lateral sclerosis functional rating scale revised.

baseline (time 1) survey and in retest (time 2) are showed in *Table 1*, and displayed an excellent internal consistency (Cronbach's alpha>0·886 for each scale). Likewise, also the test–retest reliability of ALSAQ-5 I items in the test–retest analysis (*Table 5*), was very good (Spearman's correlation coefficient>0·73 for each item).

Construct validity

Construct validity describes the extent to which the scores on a scale reflect what current theory predicts the test will show. It was examined by means of Spearman's correlations of scales for ALSAQ-40, with scales for ALS-FRS, SF-36 and FVC and muscular strength mega score (superior MS and inferior MS) (*Table 2*).

Excellent correlations in the ALSFRS-R were found between eating and drinking scale and swallowing (Rho=−0·84), between activity of daily living/independence and feeding (Rho=−0·80) and physical mobility and walking (Rho=−0·82). The five items selected (ALSAQ-5) correlated with SF-36 in the same magnitudes as the ALSAQ-40. In particular, strong correlations were found with SF-36 subscales mobility and physical functioning, and between ADL independence and vitality. All correlations are negative as high ALSAQ-40 scores correspond with low ALS-FRS and SF-36 scores.

Construct validity examined by means of Spearman's correlation of Italian ALSAQ-40 and mega score upper and lower revealed a significant correlation between physical mobility and ADL independence and mega score superior and inferior. Forced vital capacity has a significant correlation with all of ALSAQ-40 scale exclusive of emotional functioning, as shown in *Table 2*.

Relationship of subscales of ALSAQ-40 to patients' variables

To determine whether there were significant differences between the scales of Italian ALSAQ-40 and either gender or request of cognitive or motor help, a Wilcox test (also known as 'Mann–Whitney' test) was performed on the data.

No significant correlation was observed between age gender or marital status ($p>0·15$) of the subject and any of the subscale of ALSAQ, nor with cognitive or motor help required ($p>0·15$) (data not shown).

Face validity

Face validity is the need for a questionnaire to apparently tap, unambiguous and easily understood, simple by item content, an underling dimension.

Table 5 Internal reliability (Spearman's correlation) of ALSAQ-5 I: correlation of the ALSAQ-5 I in the baseline (time 1) and retest (time 2)

ALSAQ-5 I items	Time 1 versus time 2
Physical mobility	0·777
ADL/independence	0·924
Eating and drinking	0·858
Communication	0·868
Emotional functioning	0·737

Most of respondents pointed out the scale as including statements relevant to how they were feeling their condition. Even though the large patients' agreement for the questionnaire, 56·58% asked cognitive help to better understand ambiguous questions (for example, at the questions 'I have found picking things up difficult', patients asked: 'with which hand?').

Discussion

The evaluation of QoL has acquired increasing importance in degenerative diseases with poor prognosis such as ALS. The lack of treatments and the disease progression imply the need for palliative therapies to ameliorate both general health status and ultimately life quality of ALS patients. Further, protocols of ALS clinical trials usually include QoL assessment as a follow-up parameter. Most existing generic QoL instruments, as the 36-item short form health survey (SF-36)[8,25], the schedule for the evaluation of individual quality of life-direct weighting (SEIQoL-DW)[11] and the McGill quality of life questionnaire (MQOL)[12], are not suitable for ALS because they do not assess features unique to this disease. Few questionnaires have been specifically designed to evaluate QoL in ALS, namely, SIP/ALS-19[10], ALSSQOL[26] and ALSAQ[21]. SIP/ALS-19 is based on a subset of items selected by a panel of ALS specialists from the sickness impact profile (SIP), whereas amyotrophic lateral sclerosis specific QoL instrument (ALSSQOL) is derived from the McGill quality of life questionnaire (MQOL)[12] modified by changes in format and by adding questions on religiousness and spirituality. To date, only limited data are available on these two measures[27]. ALSAQ-40 is otherwise a widely used ALS patient-focused disease questionnaire[28,29] and has been already validated in Japanese[17] and Dutch language[18]. The aim of our study was to validate an Italian version of the ALSAQ-40 and its shortened form, ALSAQ-5. Italian ALSAQ-40 showed high test–retest reliability and a good construct validity, as supported by comparison with the subscales of the SF-36 and ALSFR-S. Indeed, we found a strong correlation between Italian ALSAQ-40 and ALSFRS-R as shown by comparing the score of the eating and drinking subscale with the swallowing item, and the activity of daily living/independence and feeding subscale with the physical mobility and walking item. Measures of muscle strength by MRC score and forced vitality capacity by spirometry (FVC) also correlated with the score of physical mobility and ADL/independence ALSAQ-40 scales.

Our correlations outline good construct validity and a relationship between ALSAQ-40 subscales exploring both physical functioning and motor impairment. We found no relationship between the ALSAQ-40 emotional functioning subscale and either muscle strength or functional ability. These outcomes support the theory that emotional distress and functional decline are partially independent: the relatively limited influence of health-related functional status on QoL in ALS patients has also been

shown in recent studies comparing global QoL and the ALSFRS-R[4,30,31].

Hence, ALSAQ-40 I suggests that the emotive connotations of patients assigned to their life can remain relatively positive even when their health status is severely impaired.

McDonald *et al.*[32] showed that when he compares the covariates of length of illness, severity of ALS disease and age, the relative risk of death per unit time for patients with psychological distress is 2·24 times higher than in patients with psychological well-being. The risk of dying associated with psychological distress is higher than the risk associated with increased age and is similar to that of disease severity. So, psychological status and quality of life are important prognostic factors in ALS, irrespective of length of time since diagnosis, disease severity, age and marital status as observed in our study.

The distributions of gender and disease onset in our Italian sample were similar to those of previous reports[24]. During the questionnaires, the psychologist noted if patients asked for any further help (motor help or cognitive help, to better understand the questions). An high request rate for cognitive (56·6%) and motor (32·9%) help was observed, thus suggesting that a face to face interview is a good approach for ALSAQ-40 assessment.

In conclusion, we found that the Italian ALSAQ-40 and ALSAQ-5 are valid, reliable disease-specific QoL instruments for Italian MND patients, as shown also in other language versions. The results of this study show that the Italian ALSAQ-40 and ALSAQ-5 are useful measures of health status in ALS and that their psychometric indexes are comparable to those of the original UK version.

A copy the instrument translated and validated in Italian is reproduced in Appendix.

References

1 Worms M. The epidemiology of motor neuron disease: A review of recent studies. *J Neurol Sci* 2001; **191**: 3–9

2 Jenkinson C, Swash M. Health outcomes measures. In: Brown RH, Menninger V, Swash M. *Amyotrophic Lateral Sclerosis*, London: Martin Dunitz, 2000: pp. 377–387

3 Iwasaki Y, Iguchi H, Ichikawa I, *et al.* Fatigue and depression are associated with poor quality of life in ALS. *Neurology* 2003; **61**: 827–828

4 Stromberg SF, Weiss DB. Depression and quality of life issues in patients with amyotrophic lateral sclerosis. *Curr Treat Options Neurol* 2006; **8**: 410–414

5 Chiò A, Gauthier A, Montuschi A, *et al.* A cross sectional study on determinants of quality of life in ALS. *J Neurol Neurosurg Psychiatry* 2004; **75**: 1597–1601

6 Walsh SM, Bremer B, Felgoise FH, *et al.* Religiousness is related to quality of life in patients with ALS. *Neurology* 2003; **60**: 1527–1529

7 Lee JN, Rigby SA, Burcahrdt F, *et al.* Quality of life issues in motor neuron disease: The development and validation of a coping strategies questionnaire, the MND coping scale. *J Neurol Sci* 2001; **191**: 79–81

8 Ware JE, Sherbourne CD. The MOS 36-item short form health survey 1: Conceptual framework and item selection. *Med Care* 1992; **30**: 473–483

9 Jenkinson C, Hobart J, Chandola T, *et al.* Use of the short form health survey (SF-36) in patients with amyotrophic lateral sclerosis: Tests of data quality, score reliability response rate and scaling assumptions. *J Neurol* 2002; **249**: 178–183

10 Mc Guire D, Garrison L, Armon C, *et al.* Relationship of the tufts quantitative neuromuscular exam and sickness impact profile in measuring disease progression in ALS. *Neurology* 1996; **46**: 1442–1444

11 O'Boyle CA. The schedule for the evaluation of individual quality of life (SEIQoL). *Int J Ment Health* 1994; **23**: 3–23

12 Cohen SR, Mount BM, Strobel MG, *et al.* The McGill Quality of Life Questionnaire: A measure of quality of life appropriate for people with advanced disease. A preliminary study of validity and acceptability. *Palliat Med* 1995; **9**: 2007–2019

13 Jenkinson C, Fitzpatrick R, Brennan C, *et al.* Development and validation of the ALSAQ-40. *J Neurol* 1999; **246** (Suppl. 3): S16–S21

14 Jenkinson C, Fitzpatrick R. Reduced item set for the amyotrophic lateral sclerosis assessment questionnaire: Development and validation of the ALSAQ-5. *J Neurol Neurosurg Psychiat* 2001; **70**: 70–73

15 Jenkinson C, Fitzpatrick R, Michael Swash, *et al. ALSAQ User Manual: Amyotrophic Lateral Sclerosis Assessment Questionnaire*, Oxford: Health Services Research Unit, 2001

16 Jenkinson C, Fitzpatrick R, Swash M, *et al.* Comparison of the 40-item Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40) with a short-form five-item version (ALSAQ-5) in a longitudinal survey. *Clin Rehabil* 2007; **21** (3): 266–272

17 Yamaguchi T, Ohbu S, Saito M, *et al.* Validity and clinical applicability of the Japanese version of amyotrophic lateral sclerosis-assessment questionnaire 40 (ALSAQ-40). *No To Shinkei* 2004; **6**: 483–694

18 Maessen M, Post MW, Maillé R, *et al.* Validity of the Dutch version of the Amyotrophic Lateral Sclerosis Assessment Questionnaire, ALSAQ-40, ALSAQ-5. *Amyotroph Lateral Scler* 2007; **8**: 96–100

19 Brooks BR, Miller RG, Swash M, *et al.* El Escorial Revisited: Revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Other Motor Neuron Disord* 2000; **1**: 292–299

20 Jenkinson C, Fitzpatrick R, Brennan C, *et al.* Evidence for the validity and reliability of the ALS assessment questionnaire: the ALSAQ-40. *Amyotroph Lateral Scler Other Motor Neuron Disord* 1999; **1**: 33–40

21 Cedarbaum JM, Stambler N, Malta E, *et al.* The ALSFRS-R: A revised ALS functional rating scale that incorporates assessments of respiratory function. BDNF ALS study group (Phase III). *J Neurol Sci* 1999; **169**: 13–21

22 Apolone G, Mosconi P, Ware JE. *Questionario Sullo Stato di Salute SF-36*, Milano: Guerini e Associati, 1997

23 R Development Core Team. *R: A Language and Environment for Statistical Computing*, Vienna: R Foundation for Statistical Computing, 2005, available at: http://www.R-project.org

24 Brown RH, Menninger V, Swash M. *Amyotrophic Lateral Sclerosis*, London: Martin Dunitz, 2001

25 Jenkinson C, Hobart J, Chandola T, *et al.* Use of the short form health survey (SF-36) in patients with amyotrophic lateral sclerosis: Tests of data quality, score reliability response rate and scaling assumptions. *J Neurol* 2002; **249**: 178–183

26 Simmons Z, Felgoise FH, Bremer BA, *et al.* The ALSSQOL balancing physical and nonphysical factors in assessing quality of life in ALS. *Neurology* 2006; **67**: 1659–1664

27 Damiano A, Patrick DL, Guzman GI, *et al.* Measurement of health related quality of life in patients with amyotrophic lateral sclerosis in clinical trials of new therapies. *Med Care* 1999; **37**: 15–26

28 Kiebert GM, Green C, Murphy C, *et al.* Patient's health related quality of life and utilies associated with different sates of amyotrophic lateral sclerosis. *J Neurol Sci* 2001; **191**: 87–93

29 Sancho PO, Boisson D. Handicap and quality of life evaluation in ALS. *Rev Neurol (Paris)* 2006; **162**: 4S205–4S207

30 Neudert C, Wasner M, Borasio GD. Patients' assessment of quality of life instruments: A randomised study of SIP, SF-36 and SEIQoL-DW in patients with amyotrophic lateral sclerosis. *J Neurol Sci* 2001; **191**: 103–109

31 Clarke S, Hickey A, O'Boyle C, *et al.* Assessing individual quality of life in amyotrophic lateral sclerosis. *Qual Life Res* 2001; **10**: 149–158

32 McDonald ER, Wiedenfeld SA, Hillel A, *et al.* Survival in amyotrophic lateral sclerosis: The role of psychological factors. *Arch Neurol* 1994; **51**: 17–23

APPENDIX 1: ITALIAN VERSION OF ALSAQ-40

ALSAQ-40
Amyotrophic Lateral Sclerosis Assessment Quastionnaire

Le seguenti affermazioni si riferiscono ad alcune difficoltà che lei potrebbe aver riscontrato durante le ultime due settimane. Indichi per favore, ponendo una crocetta sulla casella appropriata, come si è sentito riguardo le seguenti affermazioni.

Se lei non può compiere affatto l'azione indicata dall'affermazione, la preghiamo di segnare la casella corrispondente a: ***Sempre / Non posso farlo affatto***

Quanto spesso durante le ultime due settimane ha pensato che le seguenti affermazioni fossero vere?

Ponga per favore una crocetta nella casella corrispondente ad ogni affermazione

	Mai	Raramente	Qualche Volta	Spesso	Sempre /Non posso farlo affatto
1) Ho trovato difficile camminare anche per brevi distanze, per esempio in casa	☐	☐	☐	☐	☐
2) Sono caduto/a e mentre camminavo	☐	☐	☐	☐	☐
3) Sono inciampato/a mentre camminavo	☐	☐	☐	☐	☐
4) Ho perso l'equilibrio mentre camminavo	☐	☐	☐	☐	☐
5) Ho dovuto concentrarmi nel camminare	☐	☐	☐	☐	☐

6) Camminare mi ha stancato moltissimo	☐	☐	☐	☐	☐
7) Ho sentito male alle gambe mentre camminavo	☐	☐	☐	☐	☐
8) Ho trovato difficile salire e scendere le scale	☐	☐	☐	☐	☐
9) Ho trovato difficile stare in piedi	☐	☐	☐	☐	☐
10) Ho trovato difficile alzarmi dalle sedie	☐	☐	☐	☐	☐
11) Ho trovato difficile usare le braccia e le mani	☐	☐	☐	☐	☐
12) Ho trovato difficile girarmi e muovermi nel letto	☐	☐	☐	☐	☐
13) Ho avuto difficoltà nell'afferrare gli oggetti	☐	☐	☐	☐	☐
14) Ho trovato difficile tenere in mano libri o giornali, o girarne le pagine	☐	☐	☐	☐	☐
15) Ho trovato difficile scrivere con chiarezza	☐	☐	☐	☐	☐
16) Ho trovato difficile fare i lavori in casa	☐	☐	☐	☐	☐
17) Ho avuto difficoltà nel mangiare con le posate	☐	☐	☐	☐	☐
18) Ho avuto difficoltà nel pettinarmi o nel lavarmi i denti	☐	☐	☐	☐	☐

19) Ho trovato difficile vestirmi	☐	☐	☐	☐	☐
20) Ho trovato difficoltà nel lavarmi sul lavabo del bagno	☐	☐	☐	☐	☐
21) Ho trovato difficoltà nel deglutire	☐	☐	☐	☐	☐
22) Ho trovato difficoltà nel mangiare cibi solidi	☐	☐	☐	☐	☐
23) Ho trovato difficoltà nel bere bevande liquide	☐	☐	☐	☐	☐
24) Ho trovato difficoltà nel partecipare alle conversazioni	☐	☐	☐	☐	☐
25) Credo che non sia stato facile capirmi quando parlavo	☐	☐	☐	☐	☐
26) Ho balbettato o farfugliato mentre parlavo	☐	☐	☐	☐	☐
27) Ho dovuto parlare molto lentamente	☐	☐	☐	☐	☐
28) Ho parlato meno di quanto fossi solito/a fare	☐	☐	☐	☐	☐
29) Mi sono sentito/a frustrato/a a causa del mio modo di parlare	☐	☐	☐	☐	☐
30) Mi sono sentito/a a disagio a causa del mio modo di parlare	☐	☐	☐	☐	☐
31) Mi sono sentito/a solo/a	☐	☐	☐	☐	☐
32) Mi sono annoiato/a	☐	☐	☐	☐	☐
33) Mi sono sentito imbarazzato/a nelle situazioni sociali	☐	☐	☐	☐	☐

34) Mi sono sentito/a senza speranza per il futuro	☐	☐	☐	☐	☐
35) Ho temuto di essere un peso per gli altri	☐	☐	☐	☐	☐
36) Mi sono chiesto/a perché andare avanti	☐	☐	☐	☐	☐
37) Ho provato rabbia a causa della mia malattia	☐	☐	☐	☐	☐
38) Mi sono sentito depresso/a	☐	☐	☐	☐	☐
39) Mi sono preoccupato/a di come la malattia potrà colpirmi nel futuro	☐	☐	☐	☐	☐
40) Mi sono sentito/a privato/a della mia libertà	☐	☐	☐	☐	☐

Per favore si assicuri di aver segnato una casella per ciascuna domanda.

Grazie di aver compilato questo questionario.