

Original Article

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Unmet supportive care needs mediate the relationship between functional status and quality of life in patients with amyotrophic lateral sclerosis

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Abstract

Objective. The aims of this study were to identify the unmet care needs and to examine the mediating effect of unmet supportive care needs in the relationship between functional status and quality of life (QOL) in Korean patients with amyotrophic lateral sclerosis (ALS).

Method. This was a cross-sectional study conducted among 186 patients with ALS recruited from a tertiary hospital in Seoul, South Korea. ALS patients' functional status, unmet supportive care needs, and QOL were assessed by Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised, the Amyotrophic Lateral Sclerosis Supportive Care Needs Instrument, and the Amyotrophic Lateral Sclerosis Specific Quality of Life – Revised Instrument, respectively. Mediation analysis was tested using Baron and Kenny's regression analysis and a Sobel test.

Result. The mean score for functional status was 33.35 ± 8.89 ; for unmet supportive care needs it was 2.40 ± 0.66 ; and for QOL it was 4.95 ± 1.29 . Functional status was significantly correlated with unmet care needs and QOL. Unmet care needs satisfaction demonstrated a complete mediating effect on the relationship between functional status and QOL of the patients with ALS ($\beta = -0.53$, $p < 0.001$) and the effect was significant (Sobel test; $Z = 5.48$, $p < 0.001$).

Significance of results. Although QOL was negatively affected by the functional status in our sample, the relationship was fully mediated via unmet supportive care needs. Because there is no cure for ALS, and the condition is rapidly progressive with a lethal outcome, providing care by meeting patients' needs is a critical aspect of caring for these patients. Early assessment of supportive care needs, providing services, and referring ALS patients to appropriate resources could enhance their QOL.

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive and fatal neurodegenerative disease. Patients with ALS experience symptoms that include muscle weakness, atrophy, and spasticity, as well as bulbar symptoms such as dysphagia or dysarthria. The later stages of ALS are characterized by severely limited mobility and respiratory failure, one of the primary causes of death (Clarke & Levine, 2011). ALS is also associated with nonmotor symptoms, including pain, behavioral and cognitive deficiencies, and psychological problems, such as depression and anxiety (Fang et al., 2017). Moreover, patients may pose the risk of economic burden (Oh et al., 2015b) and family problems linked to their disease experiences (Oh et al., 2015a). In addition to these difficulties, patients afflicted with ALS experience a rapid decline in functioning (King et al., 2009), during the unrelenting progression of the disease, which negatively affects their lives. Because there is no cure for this disease, caring for patients with ALS typically involves some effort to maximize the quality of life (QOL) (Pagnini & Simmons, 2018).

There is no agreed-on definition of QOL in ALS (Burns et al., 2012). The World Health Organization defines QOL as "...a broad-ranging concept affected in a complex way by the person's physical health, psychological state, level of independence, social relationships, personal beliefs and their relationship to salient features of their environment" (WHOQOL Group, 1994). Because QOL is a subjective concept reflecting perceived wellbeing in various areas of life, it is influenced not only by health conditions but also cultural perspectives, value, and environmental supports (Simmons et al., 2006). There has been some debate on the relationship between QOL and progression of the disease in ALS patients (Shamshiri et al., 2016). Studies in Western countries have provided increasing evidence that the QOL of patients with ALS does not change over time, despite a decrease in functional status (Simmons, 2015); the worse the functional status is, the lower the QOL in non-Western

countries' studies (Geng et al., 2017; Oh et al., 2018; Shamshiri et al., 2016). The latter results could be explained by the problems associated with ALS, such as dependence on others and the negative effects on patients as the disease progresses, which restricts their normal daily activities. To preserve the optimal QOL for patients suffering from this devastating disease, it is important to provide supportive care that meets the complex and distinct needs of each patient (Oh & Kim, 2017).

ALS requires a different approach toward each patient, because of the variation in the lived individual experiences as well as the homogeneity of the clinical features, such as personal symptoms and disease stages. Furthermore, in addressing the patients' needs, consideration must be given not only to the healthcare professionals' perspectives, but also to each patient's perspective (Ang et al., 2015). Although there has been growing recognition of supportive care needs in ALS, the subject has been analyzed mostly in a qualitative way (Oh & Kim, 2017). Recently, an instrument that assesses the supportive care needs of patients with ALS has been developed (Oh et al., 2018) based on the supportive care needs framework in ALS patients, which comprises seven subdomains: physical, psychological, emotional, social, spiritual, informational, and practical needs (Oh & Kim, 2017). Supportive care is most extensively studied in cancer, and many of those studies have shown that unmet patients' care needs are associated with a higher symptom burden and lower QOL (Cheng et al., 2016; So et al., 2014). However, there is little quantitative study regarding unmet supportive care needs, especially their buffering effects between functional status and QOL, in the ALS population. Therefore, the aims of this study were to identify ALS patients' unmet care needs and to examine the mediating effect of the unmet supportive care needs in the relationship between functional status and QOL among South Korean patients with ALS.

Methods

Design

This study was a cross-sectional survey study.

Sample and data collection

A total of 190 ALS patients were recruited by convenience sampling from a tertiary hospital (Hanyang University Hospital) in Seoul, South Korea, between September 2017 and June 2018. The study sample size was calculated by G*Power 3.1.9.2 using the following parameters for the F-test in linear multiple regression: an effect size of 0.1, a significance level of 0.05, study power of 95%, two variables for predictive factors, and a 20% dropout rate. All participants were older than 20 years and were diagnosed with definite, probable, probable laboratory-supported, or possible ALS, according to the El Escorial revised criteria (Brooks et al., 2000). Participants were excluded if they had been diagnosed with frontotemporal dementia, severe depression, or schizophrenia.

Those who were willing to participate in the study were given a detailed explanation before signing an informed consent form; they were then asked to complete the self-reported Amyotrophic Lateral Sclerosis Supportive Care Needs Instrument (ALSSCN) (Oh et al., 2018) and the Amyotrophic Lateral Sclerosis Specific Quality of Life – Revised Instrument (ALSSQOL-R) (Oh et al., 2017). The patients' responses were filled in by the researcher if

they could not mark the questionnaire items because of upper extremity weakness. Also, the following data were collected from a medical chart review: demographics (age, gender, religious beliefs, education level), clinical features (site of symptom onset), and Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised (ALSFRS-R) (Cedarbaum et al., 1999).

Measurements

Functional status was measured using the ALSFRS-R, the most widely used surrogate markers of disease progression of ALS in clinical research, developed by Cedarbaum et al. (1999). The ALSFRS-R contains 12 items with three domains of bulbar (items 1–3), motor (items 4–9), and respiratory function (items 10–12) (Franchignoni et al., 2013). Each item is rated on a 5-point Likert scale (0–4), and the total score is the sum of the scores of 12 items; the higher scores indicate better functional status. In this study, the score evaluated by professionals (neurologists or coordinating nurses) was used from the medical chart review. The Cronbach's alpha was 0.89 in this study.

Unmet supportive care needs were measured using the ALSSCN developed by Oh et al. (2018). The ALSSCN is a 37-item self-reported questionnaire comprising seven subscales that assess physical, psychological, emotional, spiritual, social, informational, and practical unmet needs based on a supportive care needs framework (Oh & Kim, 2017). Each item is rated on a 4-point Likert scale (1–4), and the total score (or the subtotal scores) is the average of the scores of all 37 items (or the number of each subdomain's items); higher scores indicate higher unmet supportive care needs. The Cronbach's alpha was 0.94 in this study.

QOL was measured by the ALSSQOL-R developed by Simmons et al. (2006). ALSSQOL-R is a 46-item self-reported questionnaire that assesses six subscales (physical symptoms, bulbar function, negative emotion, interaction with people and the environment, intimacy, and religiosity). Each item is assessed on an 11-point numeric scale (0–10), and several items required transposing (items 1–10, 17, 22, 24, 25, 28, 31, and 34). The total score is an average of 46 items, and the total score varies from 0 (worst) to 10 (best). In this study, a Korean version of ALSSQOL-R, validated by Oh et al. (2017) was used. The Cronbach's alpha was 0.91 in this study.

Ethical consideration

This study was conducted with the approval of the Institutional Review Board of the hospital (HYUH 2016-04-045). All patients were informed about the study's objective and gave their informed consent.

Data analysis

SPSS 21.0 was used to analyze the data, and a two-sided *p* value < 0.05 was considered statistically significant. All variables of the participants were expressed by descriptive or frequency analysis. Pearson's correlation coefficients were calculated to examine the relationship between functional status, unmet supportive care needs, and QOL.

The mediating effects of unmet supportive care needs on functional status and QOL were analyzed using Baron and Kenny's steps for mediation (Baron & Kenny, 1986). The first step involved regressing the mediator (unmet supportive care needs) on the independent variable (functional status); in the second

step, the dependent variable (QOL) was regressed on the independent variable (functional status); and in the third step, we regressed the dependent variable (QOL) on both the independent variable (functional status) and the mediator (unmet supportive care needs). To establish mediation, the following three conditions must be met: first, the independent variable must affect the mediator; second, the independent variable must affect the dependent variable; and third, the mediator must affect the dependent variable. If these conditions all hold in the predicted direction, then the effect of the independent variable on the dependent variable must be less in the third equation than in the second. Perfect mediation holds if the independent variable has no effect when the mediator is controlled (Baron & Kenny, 1986). Finally, a Sobel test (Sobel, 1982) was performed to examine whether a mediator variable significantly carries the influence of an independent variable on a dependent variable.

Results

A total of 186 participants were analyzed, excluding four with unreliable data. The mean age of the participants was 56.73 ± 10.56 years; 58.06% of the participants were males. The mean number of years of education was 12.10 ± 3.62 , and 52.69% of participants had religious beliefs. The symptom onset site was bulbar in 15.59% of participants. The overall mean scores of functional status, unmet supportive care needs, and QOL were 33.35 ± 8.89 of 48, 2.40 ± 0.66 of 4, and 4.95 ± 1.29 of 10, respectively. The highest scored subdomain of unmet supportive care needs was informational needs (2.94 ± 0.92), followed by practical needs (2.69 ± 0.97), psychological needs (2.50 ± 0.79), emotional needs (2.33 ± 1.02), social needs (2.15 ± 0.82), physical needs (2.10 ± 0.75), and spiritual needs (2.02 ± 0.88) (Table 1).

Table 2 shows the correlations between demographics, functional status, unmet supportive care needs, and QOL. Patients who is male or has religious belief had higher QOL, and education lever positively correlated with QOL. Overall functional status was significantly negatively correlated with unmet supportive care needs ($r = -0.56, p < 0.001$), and positively correlated with QOL ($r = 0.39, p < 0.001$); and unmet supportive care needs was significantly negatively correlated with QOL ($r = -0.58, p < 0.001$).

Regression analysis conducted to verify the mediation effect of the unmet supportive care needs on the relationship between functional status and QOL is described in Table 3. In the first step, the results of the regression analysis indicate that the effect of functional status on unmet supportive care needs was significant ($\beta = -0.56, p < 0.001$), supporting functional status as a reasonable explanation for the 31% of variance in unmet supportive care needs. Also, in the second step, functional status significantly affects QOL ($\beta = 0.39, p < 0.001$), supporting functional status as a reasonable explanation for the 31% of variance in QOL. In the last step, both functional status, as an independent variable, and unmet supportive care needs, as the mediator, entered the regression model, with QOL as a dependent variable. The results indicated that only unmet supportive care needs were significant predictive variables of QOL ($\beta = -0.53, p < 0.001$). In this model, the standardized regression coefficient of functional status from 0.09, and was not significant with respect to QOL ($\beta = 0.09, p = 0.205$), indicating that the complete mediating effect of unmet supportive care needs was able to explain 33% of QOL (Table 3, Figure 1). Finally, a Sobel test found that the mediating effects of unmet supportive care needs on the relationship between functional status and QOL was significant ($Z = 5.48, p < 0.001$).

Table 1. Descriptive analysis of demographics and study variables

	Category	<i>n</i> (%) or <i>M</i> \pm <i>SD</i>	
Age, years		56.73	10.56
Gender	Female	78	41.94
	Male	108	58.06
Education, years		12.10	3.62
Religiousness	Yes	98	52.69
	None	88	47.31
Site of symptom onset	Bulbar	29	15.59
	Spinal	157	84.41
ALS duration, months		35.56	30.61
Functional status, ALSFRS-R	Overall	33.35	8.89
	Bulbar function	9.67	2.35
	Motor function	13.61	6.39
	Respiratory function	10.07	2.74
	Unmet supportive care needs, ALSSCN	Overall	2.40
	Physical	2.10	0.75
	Psychological	2.50	0.79
	Emotional	2.33	1.02
	Spiritual	2.02	0.88
	Social	2.15	0.82
	Informational	2.94	0.92
	Practical	2.69	0.97
QOL (ALSSQOL-R)		4.95	1.29

ALSFRS-R, Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised; ALSSCN, Amyotrophic Lateral Sclerosis Supportive Care Needs Instrument; ALSSQOL-R, Amyotrophic Lateral Sclerosis Specific Quality of Life - Revised Instrument.

Table 2. Correlations between functional status, unmet supportive care needs, and quality of life

	Unmet supportive care needs <i>r</i> (<i>p</i>)	Quality of life <i>r</i> (<i>p</i>)
Age	-0.02 (0.817)	-0.07 (0.336)
Gender (reference: female)	-0.13 (0.073)	0.16 (0.025)
Education	-0.09 (0.212)	0.27 (<0.001)
Religious (reference: none)	-0.05 (0.465)	0.21 (0.003)
Functional status	-0.56 (<0.001)	0.39 (<0.001)
Bulbar function	-0.40 (<0.001)	0.44 (<0.001)
Motor function	-0.54 (<0.001)	0.31 (<0.001)
Respiratory function	-0.22 (0.003)	0.16 (0.034)
Unmet supportive care needs		-0.58 (<0.001)

Discussion

To the best of our knowledge, this is the first study to analyze the relationship between unmet supportive care needs, functional status, and QOL in the ALS population. First, we found that functional status was positively correlated with QOL. Because there

Table 3. The mediation effect of unmet supportive care needs on the relationship between functional status and quality of life

Step	Independent variable	Dependent variable	B	β	t	p	Adjust R ²	F	p
Step 1	Functional status	Unmet supportive care needs	-0.04	-0.56	-9.25	<0.001	0.31	85.49	<0.001
Step 2	Functional status	Quality of life	0.06	0.39	5.72	<0.001	0.15	32.67	<0.001
Step 3	Functional status	Quality of life	0.01	0.09	1.27	0.205	0.33	46.94	<0.001
	Unmet supportive care needs	Quality of life	-1.03	-0.53	-7.22	<0.001			

Sobel test: Z = 5.48, p < 0.001.

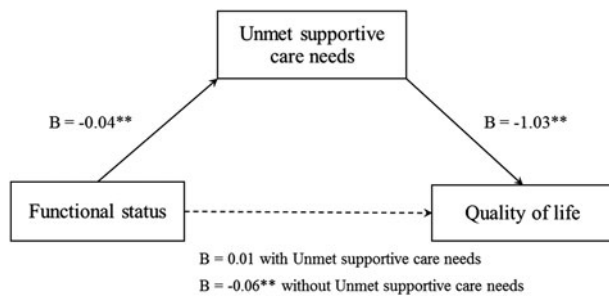


Fig. 1. Analysis of Unmet Supportive Care Needs as a Mediator of the Relation between Functional Status and Quality of Life. **p < .01.

has been no biomarker for tracking diseases severity in ALS, functional status is usually used as a primary outcome in clinical trials because it is a feature of disease progression (Paganoni et al., 2014). Although a number of studies have examined the relationship between the progress of this disease and QOL, such studies have produced inconsistent results concerning such relationship (Shamshiri et al., 2016). Many recent studies have indicated that QOL in patients with ALS appears to be maintained over time as physical function declines and is not equivalent to their disease progression (Felgoise et al., 2018), which can be explained by the results of recalibrating their expectations, the so-called “response shift” (Simmons, 2015). However, other studies show that QOL worsens as the disease progresses. One study conducted among patients from South Korea (Oh et al., 2017) and other non-Western countries (Geng et al., 2017; Shamshiri et al., 2016) found a correlation with a medium effect size between functional status and QOL, which is in line with this study.

Second, functional status was associated with unmet supportive care needs. As functional status deteriorated, more complex and more intensive care needs were required in this population. Although this result is intuitive, to the best of our knowledge, the present study is the first to document and quantify this relationship. This is probably because the worse the patient’s functional impairment, the more likely he or she will be to face complex difficulties. These include not only the deterioration of motor function, but also associated psychosocial problems such as social isolation, depression, economic difficulties (Oh et al., 2015b), and non-motor symptoms (fatigue, pain, and sialorrhea and sleep disturbance) (Fang et al., 2017). Also, these results are consistent with previous studies that explored the relationship among patients with cancer and other neurological diseases (Saleem et al., 2013; Sandgren et al., 2010). Supportive care needs have been extensively studied in cancer care. Cancer patients generally experience a relatively long period of high functioning, followed by a short period of rapid decline toward death, but with ALS, there is a prolonged period of frailty and disability (Ang et al., 2015). Also, to date, no cure has been found for ALS,

whereas generally cancer can be treated in several ways, including surgery and chemotherapy. The rareness of ALS (approximately 1.75 per 100,000 people) (Marin et al., 2017) is likely to make patients face greater challenges and experience multiple unmet needs as the disease progresses (von der Lippe et al., 2017). Such findings emphasize the need for further research investigating unmet supportive care needs and the intrinsic and external factors affecting ALS patients.

Third, unmet supportive care needs were negatively correlated with QOL. Moreover, it supports the total mediation role of affect in explaining the relationship between functional status and QOL, which is in line with findings in many studies among cancer patients (Cheng et al., 2016; So et al., 2014). In other words, this implies that the QOL of patients with ALS, whose physical function is declining, can be maintained when their supportive care needs are satisfied. This finding is important because physical deterioration in ALS is not reversible, and therefore, enhancing the QOL of patients is essential in caring for patients with ALS. In addition, our findings indicate that Korean ALS patients have substantial supportive care needs that may be insufficient. Although Korea is a high-income country with a relatively well-established health care system, this system does not fully meet the patients’ needs (Molassiotis et al., 2017). Recent cross-country studies have revealed that Korean cancer patients have higher unmet needs than patients from Japan, Australia, and India (Molassiotis et al., 2017). Moreover, the care services for patients with rare diseases are even more inadequate and are focused mostly on reducing patient’s medical care costs. Accordingly, such services do not play a role in the overall social safety net established for the patients (Chong & Heo, 2015). In other words, the policy does not reflect the specificity of the disease, and so the resulting burden of caring for patients is borne by family members. As a first step, identifying and addressing such needs is likely to prevent patients’ deterioration of QOL in a thorough multidisciplinary approach. Also, care in ALS is most effective when coordinated among a specialized multidisciplinary team, the community-based sector (including primary care), and palliative care teams (Hogden et al., 2017). Our study clearly emphasizes the importance of incorporating supportive care services into community care as well as routine ALS care in hospitals to satisfy patients’ complex practical care needs, including the presence of informal caregivers and transportation. In particular, our findings show that among the highest unmet needs are informational needs, which suggests the importance of offering more information about ALS and ALS related self-care.

This study has the following limitations for generalization. First, the cross-sectional design does not allow a definitive conclusion for causation to be drawn. Second, all of the patients were recruited from a single tertiary referral center, which may lead to selective bias. A further multicenter longitudinal study with a larger sample may help to verify our study. Nevertheless, this study first examines

unmet supportive care needs and their relationship with functional status and QOL in ALS patients. Also, because we use disease-specific, validated, and psychometrically sound measurements, validity could be higher. However, this mediating effect should be supported not only by further research with repeated longitudinal design studies, but also studies from other countries because Korea has a different health care social welfare system. Also, other variables that can affect QOL (e.g., mood status, social supports) should be included in further studies.

Although QOL is negatively affected by functional status in our sample, the relationship was fully mediated via unmet supportive care needs. Because there is no cure for ALS, and the illness progresses rapidly with a lethal outcome, providing care and meeting the patients' needs are critical aspects of caring for them. Through early assessment of supportive care needs, providing services, and referring patients to appropriate resources could enhance their QOL. Relatively high scores of unmet supportive care needs in our study also indicate that gaps may exist between patients' expectations and their actual experience of services. To meet the patients' complex needs, it is necessary to develop educational programs for patients, adopt a multidisciplinary approach in a hospital setting, and improve the delivery system of community services.

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