

# A Phenomenological Analysis on Quality of Life of Patients with Amyotrophic Lateral Sclerosis (ALS) in Taiwan

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## Abstract

Amyotrophic Lateral Sclerosis (ALS) is a severe and lethal progressive neurodegenerative disease, and patients with ALS are commonly called “gradually frozen people.” The purpose of this study is to investigate the influence of oropharyngeal dysphagia of patients with ALS on their quality of life. This study enrolled a total of 52 subjects (31 male subjects and 21 subjects). For measurement tools, this study used Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40) for self-assessment of patients with ALS and Questionnaire on Swallowing and Quality of Life (SWAL-QOL) to perform investigations. The results of analysis on correlation between basic information and dysphagia showed that, 4 items: “gender,” “age,” “marital status,” and “educational background” were not significantly correlated with level of dysphagia (P value >0.05). The analysis on correlation between physical symptoms and level of dysphagia showed that, 4 items: “difficulty in eating,” “incomprehensible talking,” “stutter,” “talking slowly” were significantly correlated with level of dysphagia (P value <0.05). The analysis on correlation between psychological symptoms and level of dysphagia showed that, 8 items: “talking less,” “talking depressively,” “talking shyly,” “feeling lonely,” “feeling embarrassed and stressed in public space,” “feeling hopeless about the future,” and “feeling angry about disease” were significantly correlated with level of dysphagia (P value <0.05). According to the research results, oropharyngeal dysphagia is a common symptom in patients with ALS, and it affects the quality of life of patients, especially psychological health and social life.

**Keywords:** Amyotrophic lateral sclerosis (ALS); Dysphagia; Trismus

## Introduction

Amyotrophic Lateral Sclerosis (ALS) is a progressive neurodegenerative disease involving upper motor neurons (UMN) and Lower Motor Neurons (LMN). The initial symptoms may vary with individuals. Typical symptoms are found in limbs (disability in arms and legs) and medulla oblongata (dysarthria and

dysphagia) [1]. ALS is featured by the loss of upper and lower motor neurons, including primary motor cortex, brain stem and spinal cord. The most common symptom of ALS is dysphagia. Weight loss is also a common phenomenon in patients with ALS. A cohort study on ALS, 56.3% of the patients experienced weight loss [2]. However, specific cause of ALS is still unknown [3]. Dysphagia is oropharyngeal muscle weakness caused by dysfunction, and is also the major clinical sign and problem of oropharynx of patients with ALS. Dysphagia will further lead to the risks of aspiration

pneumonia, malnutrition, and dehydration in patients with ALS [4]. Previous studies also indicated that, relevant factors leading to oropharyngeal dysphagia will affect the quality of life of patients with ALS. Patients with ALS tend to experience tongue bleeding, broken teeth, and loosening of dentures and braces due to trismus-induced excessive force and dentition dislocation. Moreover, ALS will lead to difficulty in opening and closing of mouth. Trismus will lead to limited mouth opening. In general, when the maximum mouth opening is less than 18-20 mm, it is difficult for patients to eat anything. The purpose of this study is to investigate the influence of oropharyngeal dysphagia on the quality of life of patients with ALS in Taiwan.

## Materials and Methods

This study was approved by Institutional Review Board (IRB) of Chung Shan Medical University Hospital (CSMUH) (Case No. CS13140). Questionnaires were sent to 230 patients with ALS in Taiwan via mail, and 52 valid questionnaires (31 male and 21 female patients) were returned, with a response rate of 22.60%.

For assessing quality of life of patients with ALS, this study used Questionnaire on Swallowing and Quality of Life (SWAL-QOL) and Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40) for self-assessment of patients with ALS to perform assessments. Questionnaire on Swallowing and Quality of Life (SWAL-QOL) is designed for patients with oropharyngeal dysphagia, and its purpose is to understand the influence of dysphagia on quality of life [5]. The questionnaire was completed mainly by the patients. For patients whose disease progressed to advanced stage and were unable to engage in activities and answer questions, social workers, interviewers or family members assisted patients in completing the questionnaire. Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40) is an effective fundamental scale for assessing the daily physiological functions of patients with ALS, as well as a scale for assessing disease-specific features [6]. The questionnaire was divided into 4 parts: basic information, physical parameters, psychological parameters, and swallowing parameters to systematically collect and describe the possible difficulties and conditions currently encountered by patients. Patients are asked to indicate the frequency of each event by selecting one of 5 options (Likert scale): never/rarely/sometimes/often/always or cannot do at all. Except that "never" was granted 0 point, 5 points were granted to each option, with a total score of 20 points. The higher the score was, the higher the level of dysphagia was; the lower the score was, the lower the level of dysphagia was. In this study, the threshold value of dysphagia was set at  $\geq 10$  points, while that of non-dysphagia was set at  $< 10$  points.

For the basic information of the subjects, gender was divided into "male" and "female," age was divided into young adults (30-39, 40-49, and 50-59 years old) and the elderly (60-69 and 70-79 years old), marriage status was divided into two groups - married & unmarried, and divorced & widowed, educational background was divided into two groups - "low educational background" (illiteracy, literacy but without schooling, elementary school, junior high school, and senior high school) and "high educational background" (junior college, bachelor, master, and PhD).

The variables of physical parameters were divided into 15 items: short-distance walking, fall during walking, loss of balance during walking, leg pain during walking, difficulty in climbing upstairs and downstairs, difficulty in talking, difficulty in turning over on bed, difficulty in grabbing things, difficulty in writing, difficulty in eating, difficulty in combing hair and brushing teeth, difficulty in wearing clothes, incomprehensible talking, stutter, and talking slowly.

The variables of physical symptoms were divided into 15 items: talking less, talking depressively, talking shyly, feeling lonely, feeling bored, feeling embarrassed and stressed in public space, feeling hopeless about the future, worrying about being a burden to others, rejecting the closeness and assistance of family members or others, feeling angry about disease, feeling depressed, worrying that disease condition may undermine the future, feeling no freedom, fearing the crowd, and having suicidal thought. In statistical analysis, this study used student's t-test to compare the difference in mean of the two groups. For categorical data, this study used chi-square test or Fisher exact test to test their correlation. All analyses were made using SAS 9.3 software (SAS Institute, Cary, NC).

## Results

### Analysis on Correlation between Basic Information and Level of Dysphagia

Although all demographic variables (gender, age, marital status, and educational level) were not significantly correlated with level of dysphagia ( $P$  value  $> 0.05$ ), most of the subjects experienced dysphagia ( $n=43$ ) (Table 1). According to the research results, the average age of young adults under the age of 60 who experienced dysphagia was 49.7 years old (25 patients), while that of those who did not experience dysphagia was 48.5 years old (6 patients). The average age of the elderly over the age of 60 who experienced dysphagia was 65.6 years old (18 patients), while that of those who did not experience dysphagia was 63 years old (3 patients). In addition, female patients were outnumbered by male ones, and male patients accounted for approximately 2/3 of all of the patients

Variables	With dysphagia ( $\geq 10$ ) (N=43)	Without dysphagia ( $< 10$ ) (n=9)	p-value
Gender n (%)			0.31
Male	27 (87.1)	4(12.9)	
Female	16 (30.8)	5(9.6)	
Age (mean $\pm$ SD) <sup>a</sup>			
Young	49.7 $\pm$ 7.7(n=25)	48.5 $\pm$ 9.7(n=6)	0.751
Elderly	65.6 $\pm$ 4.4(n=18)	63.0 $\pm$ 4.4(n=3)	0.354
Marital status n (%)			
Married/unmarried	39 (75.0)	8(15.4)	0.87
Divorced/widowed	4 (7.7)	1(1.9)	
Educational level			
n (%) <sup>b</sup>	24 (46.2)	4(7.7)	0.39
Low	19 (36.5)	5(9.6)	

\* a: Young: 30-39, 40-49, 50-59, Elderly: 60-69, 70-79

\* b: Low educational level: illiteracy, literacy but without schooling, elementary school, junior high school, and senior high school; high educational background: junior college, bachelor, master, and PhD.

**Table 1:** Analysis on Correlation between Research Subjects' Basic Information and Level of Dysphagia (sample size=52).

### Analysis on Correlation between Physical Parameters and Level of Dysphagia

The scores were divided into two groups: low scores (never, seldom, and sometimes) and high scores (frequently and always/completely not). According to the results of analysis on correlation between physical parameters and level of dysphagia in Table 2, difficulty in eating, incomprehensible talking, stutter, and talking slowly were significantly correlated with level of dysphagia (P value  $<0.05$ ), and the difference reached statistical significance. Among 37 patients experiencing difficulty in eating (High score), 34 (92%) experienced dysphagia. Most of the patients displaying dysphagia were incomprehensible talking (97%), stutter (96%), and talking slowly (97%).

Variables	With dysphagia ( $\geq 10$ ) (n=43)	Without dysphagia ( $< 10$ ) (n=9)	p-value
Short-distance walking			0.40
Low score	10(19.2)	3(5.8)	
High score	33(63.5)	6(11.5)	
Fall during walking			0.83
Low score	11(21.2)	2(3.9)	
High score	32(61.5)	7(13.5)	
Loss of balance during walking			0.29
Low score	8 (15.4)	1(1.9)	
High score	35(67.3)	8(15.4)	
Leg pain during walking			0.42
Low score	11(21.2)	4(7.7)	
High score	32(61.5)	5(9.6)	
Difficulty in climbing upstairs and downstairs			0.59
Low score	50(9.6)	2(3.9)	
High score	38(73.1)	7(13.5)	
Difficulty in talking			0.67

Low score	9 (17.3)	1(1.9)	
High score	34(65.4)	8(15.4)	
Difficulty in turning over on bed			0.17
Low score	6 (11.5)	3(5.8)	
High score	37(71.1)	6(11.5)	
Difficulty in grabbing			0.13
Low score	50(9.6)	3(5.8)	
High score	38(73.1)	6(11.5)	
Difficulty in writing			0.06
Low score	50(9.6)	4(7.7)	
High score	38(73.1)	5(9.6)	
Difficulty in eating			0.01
Low score	9 (17.3)	6(11.5)	
High score	34(92.0)	3(8.0)	
Difficulty in combing hair and brushing teeth			0.61
Low score	6 (11.5)	2(3.9)	
High score	37(71.2)	7(13.5)	
Difficulty in wearing clothes			0.13
Low score	50(9.6)	3(5.8)	
High score	38(73.1)	6(11.5)	
Incomprehensible talking			0.00
Low score	17(32.7)	8(15.4)	
High score	26(97.0)	1 (3.0)	
Stutter			0,03
Low score	20(38.5)	8(15.4)	
High score	23(96.0)	1(4.0)	
Talking slowly			0.00
Low score	12(23.1)	8(15.4)	
High score	31(97.0)	1(3.0)	

\* a: low scores: never, seldom, sometimes

\* b: high score: frequently, always/completely not

**Table 2:** Analysis on Correlation between Research Subjects' Physical Parameters and Level of Dysphagia (sample size=52).

### Analysis on Correlation between Psychological Parameters and Level of Dysphagia

As shown in Table 3, talking less, talking depressively, talking shyly, feeling lonely, feeling bored, feeling embarrassed and stressed in public space, feeling hopeless about the future, and feeling angry about disease were significantly correlated with dysphagia (P value<0.05). Among these variables, except for feeling hopeless about the future, most of the patients with high score suffered from dysphagia (> 95%). Among 35 patients feeling hopeless about the future, only 32 (91%) experienced dysphagia.

Variables	With dysphagia ( $\geq 10$ ) (n=43)	Without dysphagia ( $<10$ ) (n=9)	p-value
Talking less			0.01
Low score	15(65.2)	8(34.8)	
High score	28(96.6)	1(3.4)	
Talking depressively			0.02
Low score	18(34.6)	8(15.4)	
High score	25(96.2)	1(3.8)	
Talking shyly			0.02
Low score	25(48.1)	9(17.3)	
High score	18(100)	0(0)	
Feeling lonely			0.02

Low score	18(34.6)	8(15.4)	
High score	25(96.2)	1(3.8)	
Feeling bored			0.02
Low score	19(36.5)	8(15.4)	
High score	24(96.0)	1(4.0)	
Feeling embarrassed and stressed in public space			0.03
Low score	21(40.1)	8(15.4)	
High score	22(95.7)	1(4.3)	
Feeling hopeless about the future			0.04
Low score	11(21.2)	6(11.5)	
High score	32(61.5)	3(5.8)	
Worrying about being a burden to others			0.33
Low score	12(23.1)	4(7.7)	
High score	31(91.4)	5(8.6)	
Rejecting the closeness and assistance of family members or others			0.82
Low score	37(71.2)	8(15.4)	
High score	6 (11.5)	1(1.9)	
Feeling angry about disease			0.02
Low score	26(50.0)	9(17.3)	
High score	17(100)	0(0)	
Feeling depressed			0.07
Low score	23(44.2)	8(15.4)	
High score	20(38.5)	1(1.9)	
Worrying that disease condition may undermine the future			0.05
Low score	9 (17.3)	5(9.6)	
High score	34(65.4)	4(7.7)	
Feeling no freedom			0.20
Low score	9(17.3)	4(7.7)	
High score	34(65.4)	5(09.6)	
fearing the crowd			0.14
Low score	26(50.0)	8(15.4)	
High score	17(32.7)	1(1.9)	
Having suicidal thought			0.09
Low score	29(55.8)	9(17.3)	
High score	14(26.9)	0(0.0)	

\* a: low scores: never, seldom, sometimes  
 \* b: high score: frequently, always/completely not

**Table 3:** Analysis on Correlation between Psychological Parameters of Research Subjects and Level of Dysphagia (sample size=52).

## Discussion

As shown in Table 1, “age” was particularly worthy of investigation. The research subjects in this study were mainly patients with ALS. Although age was included in the correlational analysis (Table 1), dysphagia is induced according to “course of disease,” instead of “increase of age.” Therefore, this result showed that age was not significantly correlated with dysphagia (P value >0.05). Past studies showed that ALS is the most common motor neuron disease in adults [7]. The risk factors include radiation, lead poisoning, and lightning damage. The symptoms of patients with susceptibility genes are more similar to those of sporadic ALS [7]. Rehabilitation specialists indicated that, dysphagia is very common in the elderly. The degeneration of the central and peripheral sensory motor systems caused by ageing will lead to dysphagia or aggravate the existing swallowing problems [8].

The results of correlation between physical symptoms and level of dysphagia in this study showed that, 4 variables: difficulty in eating, incomprehensible talking, stutter, and talking slowly were significantly correlated with level of dysphagia, suggesting that patients experienced the phenomenon of oropharyngeal dysphagia according to the course of disease (Table 2). The past studies showed that, patients start to experience onset from bulbar muscle paralysis, and they experience the symptoms of difficulties in swallowing and talking when the movements of their limbs are still fine. Afterwards, their disease progressed to respiratory failure, and they cannot take care of daily life and engage in activities, which indirectly affects quality of life [9]. Dysphagia is associated with mouth, tongue, pharynx muscle weakness. Therefore, patients experiencing Bulbar Onset (BO) suffer from dysphagia earlier than those experiencing Spinal Onset (SO), and their suffering at advanced stage of the disease are significantly higher than that of those experiencing SO [10].

Moreover, excessive saliva is also a factor confusing patients with ALS and affecting their quality of life. Relevant studies showed that, ALS is a disease that will damage all of the voluntary muscles. In early stage, patients may also experience symptoms from medulla oblongata. Therefore, the salivation (excessive saliva) in certain patients may be a severe problem [11]. In terms of saliva, the amount of saliva secreted by patients every day is 0.5-1.5 liters. Therefore, patients find it extremely difficult and inconvenient to deal with saliva cleaning, dysphagia, and salivation every day [12]. For the excessive saliva of patients with ALS, some studies indicated that radiotherapy can be used to effectively reduce salivation [13]. The dysphagia and salivation of patients with ALS should be managed by professionals to improve their quality of life [14].

The results of correlation between psychological symptoms and level of dysphagia showed that, 8 items: talking less, talking depressively, talking shyly, feeling lonely, feeling bored, feeling

embarrassed and stressed in public space, feeling hopeless about the future, and feeling angry about disease were significantly correlated with level of dysphagia (P value <0.05), suggesting that the psychological symptoms and stress of patients caused by disease may also lead to the difficulty in chewing food during eating and cough during swallowing, namely, the so-called situation of “failure to swallow food during eating” (Table 3). According to clinical psychiatrists, psychological stress will also affect physical change and even various immune systems to make people become sick, further induce symptoms, such as physical coordination or balance disorders, paralysis or partial muscle weakness, dysphagia or sense of foreign body sensation in throat, speechlessness, and urinary retention, a.k.a., pseudo-neurological symptoms [15]. As shown in Table 3, some unexpected phenomena were particularly discovered: 7 items, worrying about being a burden to others, rejecting the closeness and assistance of family members or others, feeling depressed, worrying that disease condition may undermine the future, feeling no freedom, fearing the crowd, and having suicidal thought, were not significantly correlated with level of dysphagia. Such a phenomenon is different from the psychological status after experiencing severe disease as perceived by general people.

The patients with ALS in this study seemed to calmly face their own illness. According to the environment of patients and care quality of caregivers, if caregivers properly took care of patients with ALS during caregiving process, the negative emotions of patients with ALS could be reduced and their positive attitude towards the disease could be further improved to obtain psychological adjustment and change in concepts. Relevant studies showed that, suffering from depression in 1/5 of patients with ALS was significantly associated with the psychological factor of depression [16]. In this study, the patients also mentioned that, due to long-term sitting or lying, they experienced weak bowel movement and difficulty in defecation. Intestinal gas might also be blocked by feces and could not be discharged, which further caused bloating. Therefore, they followed medical instructions to take anti-flatulence drugs, such as Gascon, to reduce discomforts. However, this drug will lead to frequent farting. Therefore, the patients had to move around their hips to avoid the embarrassment and perplex caused by the sound and smell of farting. The patients with ALS indicated that the said situation reduced their willingness to dine in public space and engage in social activities, which also indirectly affected their social life. Relevant studies showed that, flatulence is a very common gastrointestinal disorder which has a significant influence on daily life, work, and interpersonal relationship [17].

## Research Limitations

The possible research limitations of this study are explained as follows: if social workers or family members assisting the patients in completing the questionnaire failed to stay neutral and used their subjective and autonomous perception to affect

patients, the answers might be biased and deficient in objectivity. Because the cognitive function of the patients declined, they might not understand the intention or purpose of questions and needed professional assistance and explanations. However, the patients might not be provided with professional explanations in a timely manner because professionals were not at the scene. Moreover, the factors, such as care quality of caregivers, economic status of patients and actual survival rate, and small sample size caused by rare disease, all might affect the patients' willingness to complete questionnaire, questionnaire return rate, and research results.

## Conclusion

Oropharyngeal dysphagia induced by disease in patients with ALS will affect patients' quality of life, psychological health, and social life, especially psychological health. The research results also showed that oropharyngeal dysphagia induced by disease in patients with ALS will also affect the quality of life and social life of patients with ALS.

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