

## Pain at end-of-life among patients with amyotrophic lateral sclerosis receiving noninvasive ventilation

Mitsuko Ushikubo<sup>1</sup>, Noriko Tsukagoshi<sup>1</sup>, Midori Shimizu<sup>2</sup>, Yoko Takahashi<sup>2</sup>

<sup>1</sup>Graduate School of Health Sciences, Gunma University, Japan

<sup>2</sup>Mihara Memorial Hospital Department of Nursing, Japan

DOI: <http://dx.doi.org/10.15520/ijnd.2017.vol7.iss7.233.36-39>

**Abstract:** The purpose of this study was to clarify aspects of pain before death among patients with amyotrophic lateral sclerosis (ALS) receiving noninvasive positive pressure ventilation (NPPV). The participant in this study was an excellent nurse manager. We conducted a semi-structured interview regarding three ALS patients who had been receiving NPPV during the last month before death. Data were analyzed using inductive qualitative analysis.

The deceased ALS patients who had been receiving NPPV were two males and one female ranging in age between their 60s and 70s. The period from onset of ALS to death ranged from 18–31 months. The duration of NPPV use ranged from 1.5 months to 2 years. The following seven categories were extracted as aspects of patients' pain: physical pain; anxiety about their children; regrets about life; pain from a bad family relationship; pain from strong pride; pain from fluctuating decision making about medical treatment for respiratory problems; and pain from not being able to express gratitude to others. Nurses and other health care professions should employ the concept of "Total Pain" and a community-integrated approach to provide palliative care to ALS patients receiving NPPV at end-of-life.

**Key words:** ALS, NPPV (NIV), end-of-life, pain, palliative care.

### INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease with no known cause and no effective treatment. Patients with ALS experience progressive loss of motor neurons, muscle atrophy, and increased difficulty in activities of daily life. The predominant cause of death among patients with ALS is respiratory failure.<sup>1</sup>

Recently, the use of noninvasive positive pressure ventilation (NPPV or NIV) has become increasingly common as a symptomatic therapy for respiratory failure associated with ALS and other motor neuron diseases (MND). NIV can help improve prognosis and palliate respiratory difficulties.<sup>2</sup> In addition, NIV allows some delay in the decision of whether to administer tracheostomy positive pressure ventilation (TPPV).

However, when NIV is required around the clock to alleviate dyspnea due to the progress of respiratory failure, it is considered more of a life-sustaining treatment than a palliative measure. In order to palliate respiratory distress among patients with ALS in such cases, NPPV should be discontinued and replaced with tracheostomy or TPPV. In a Japanese study by Tagami et al., 15 of 37 NIV patients had undergone tracheostomy/TPPV (rate of transition from NIV to TPPV: 41%),<sup>3</sup> and Kurihara et al. reported that approximately 50% of all ALS patients receiving NPPV in their center refused to change to TPPV.<sup>4</sup>

Although an increasing number of patients have begun

receiving NPPV,<sup>5</sup> palliative care for the patients with ALS who decline TPPV has not yet been developed.<sup>6</sup> The review article regarding palliative care at the end-of-life among people with ALS under NIV mentioned that little attention has been paid to palliate various pains for patients with ALS patients who decided NPPV as maximum medical treatment to respiratory failure.<sup>7</sup>

In the previous studies dealt with pain in the ALS or MND, only two articles described pain influences their quality of life.<sup>8,9</sup> One article discussed pain from the psychological perspective.<sup>10</sup> Most of the articles discussed about only physical pain.<sup>11</sup> However, many articles pointed out that pain in ALS or MND has under-recognized and underestimated.<sup>12,13</sup>

This study focused on end-of-life care involving the use of NIV. The aim of this study was to clarify the aspects of pain experienced by patients with ALS receiving NIV at end-of-life.

### METHODS

#### *Study design:*

This was a retrospective and qualitative-descriptive study.

#### *Participant:*

The participant in this study was a nurse manager from the hospital M specialized in neurology. She acted as the unit administrator for patients with ALS. The reason why we selected a nurse manager was that she was considered capable of taking an objective view of the nursing staff,

patients, and their families because she had substantial experience in nursing and her personal life. The nurse managers in Japan usually involve patients and their family although they don't provide nursing treatment as staff nurses do.

Another reason why we selected this participant of this study is that she is one of the rare and intractable illnesses nurses accredited by Japan Intractable Illness Nursing Society. In addition, many colleagues in neurological nursing have admitted she is an outstanding nurse. Since this study deals with the difficult and sensitive matter, we think there is no one else but her to collect rich and valuable data.

#### **Data collection:**

We conducted a semi-structured interview regarding ALS patients who had been receiving NPPV before their death at the hospital M during a 3-year period from 201X. Informed consent was obtained from the families of three patients with ALS who met these inclusion criteria. We asked the participant to explain the demographic characteristics of each patient and talk freely about how she felt regarding the pain of each ALS patient in the final month before their death.

#### **Data analysis:**

Interview data were recorded and transcribed verbatim, and then subjected to qualitative descriptive analysis. The following procedures were used for the inductive analysis of qualitative data. Three researchers independently and repeatedly read the nurse manager's responses verbatim and extracted segments in the context of one meaning, and then shortened them into two steps while keeping the same meaning. Next, after assessing the similarities and differences between each shortened segment, three researchers scrutinized, discussed, and extracted relevant categories.

To ensure trustworthiness, the results were checked by the participant in accordance with the method of member checking to ensure the trustworthiness of the qualitative analysis.

We also performed the expert checking and obtained the agreement from three nursing experts specialized in research of neuro-intractable diseases.

#### **Ethical considerations:**

The protocol of this study was approved by the ethics review committee of the hospital M (approval number 068-04). The researcher provided oral and written explanations to both the participant and the families of the deceased. The main content of the explanations included the study objectives and methods, the privacy protection policies, the voluntary nature of their participation, and the fact that they could withdraw at any time. Written consent was obtained from the participant and the family of the deceased.

## **RESULTS**

### ***Characteristics of patients with ALS:***

The deceased ALS patients who had been receiving NPPV to manage their respiratory problems comprised two males and one female ranging in age between their 60s and 70s. The period from onset of ALS to death ranged from 18–31 months, and the period of NPPV use ranged from 1.5 months to 2 years.

### ***Aspects of pain:***

Seven categories were extracted from the narratives of the nurse manager. Each category was explained using shortened sentences or segments.

#### ***Physical pain:***

The nurse manager felt that all ALS patients had suffered physical pain.

*"I understood her painful situation. I felt that her physical deterioration and fatigue in the last month was causing her a lot of pain."* (for Patient A)

*"His disease progression was so rapid. I guessed that this rapid progression truly afflicted him with pain."* (for Patient B)

*"I felt that he was suffering from intense physical pain, but he suppressed it with his tenacious courage."* (for Patient C)

#### ***Anxiety about their children:***

*"I thought that her biggest source of pain was concern about her disabled son."* (for Patient A)

*"His son had heart disease. His wife told him that his son hovered between life and death during his hospitalization. I guessed that it was very painful for him that he could not do anything for his sick son."* (for Patient C)

#### ***Regrets about life:***

This category was extracted from the narrative about Patient A only. The nurse manager knew the background of this patient very well.

*"She liked her job and was active and hard working. I understood that she wished she could work a little bit longer. She wished to work until retirement."* (for Patient A)

#### ***Pain from a bad family relationship:***

This category was extracted from the narrative about Patient B only. The nurse manager knew the patient's family background and relationships very well.

*"He wanted his family to understand his pain and give gentle consideration to him. But his wife could not empathize or understand him very well. This made him angry. I thought this situation was very painful and difficult for him."* (for Patient B)

### **Pain from strong pride:**

This category was extracted from the narrative about Patient B only. The nurse manager understood his character very well.

*“He had strong pride and wanted to live up to his ideal. That was why he could not complain that the situation was too painful. That was too difficult for him.”* (for Patient B)

### **Pain from fluctuating decision making about medical treatment for respiratory problems:**

The nurse manager provided long and abundant patient narratives, which suggests that she perceived a great deal of pain regarding decision making in respiratory medical treatment. Several examples are as follows:

*“I thought that she felt sorry about causing her husband a lot of grief if she decided not to undergo TPPV. Her husband wanted her to undergo TPPV and live with him as long as possible, but he understood her thoughts and worried that he would not be able to handle the caregiving burden, because their son was also disabled. My heart ached for her husband’s pain, which resulted from the conflict between his wishes and her true intentions..”* (for Patient A)

*“He declined TPPV, but his wife didn’t. She wanted him to live with the respirator. So, I felt his intense mental pain stemming from a feeling of crisis that he might be connected to a respirator against his will.”* (for Patient C)

### **Pain from not being able to express gratitude to others:**

This category was extracted from the narrative about Patient C. The nurse manager understood his character very well and gave thorough consideration to his situation.

*“He was polite. Every time he was treated, he expressed his gratitude to the nursing staff, even though he was in tremendous pain. Near the end-of-life, he required nursing care in all aspects of his daily life. However, he could no longer communicate anything. There was no way for him to express his gratitude. I felt that he was experiencing intense pain.”* (for Patient C)

## **DISCUSSION**

The patients’ pain extracted from the narratives of the participant included all aspects of total pain: physical; psychological; social; and spiritual pain. The participant had a deep understanding of the pain of each ALS patient and held highly person-centered views of the patients’ characteristics and their family backgrounds.

It is extremely difficult to provide palliative care for physical pain to patients with ALS receiving NPPV at the end-of-life. A previous study clarified physical pain experienced during the last month before death.<sup>14</sup> Staff nurses are extremely busy to provide care peculiar to

NPPV treatment and tend to palliate only physical pain, because it is difficult for them to adjust NPPV pressure in order to palliate progressive respiratory distress, care for facial skin breakdown with NPPV masks, provide oral care and sputum suctioning without taking the patient’s ventilator mask off for more than a few seconds, and palliate intolerable body pain. Although morphine (opioids) is typically used, it sometimes fails to palliate the pain. The participant in this study also recognized the physical pain of each patient.

In addition, the participant felt the pain experienced by the patients and their families from fluctuating decision making about medical treatment for respiratory problems. She understood how this pain fluctuated based on whether the patients received tracheostomy or the use of a ventilator and its association with the condition of the familial relationship. These findings suggest that nurses and other health care professions need to understand patients’ feelings and thought processes regarding decision making in order to help provide death with dignity.

Patients living with ALS experience a variety of complicated problems from the early stage of their illness until death. Therefore, it is important to apply the notion of “Total Pain” to patients with ALS, similar to patients with cancer in palliative care.<sup>15,16</sup> The conditions of patients with ALS gradually aggravates and becomes increasingly difficult to communicate their thoughts and feelings even though their conscientiousness is intact. Therefore, nurses should try to understand not only physical pain, but also the thoughts and feelings of patients until their death.

The patients with ALS may die in their home, at a care house, in a respite care facility or in the hospital. It is hard to predict the time of approaching death of patients with ALS; they are not uncommon to die suddenly.<sup>17</sup> The nurses and other professions need to practice community-based integrated care and share pain and thoughts of ALS patients. Network care is associated with reduced hospital administration, reduced deterioration of function, and delayed mortality in patients with ALS.<sup>18</sup> Moreover, an integrated approach that coordinates care between the hospital and other care settings in community can influence the palliation of dying and the quality of death of patients with ALS.

## **CONCLUSION**

The following seven categories were extracted as aspects of pain among deceased patients with ALS who had been receiving NPPV based on the perspective of a nurse manager: physical pain; anxiety about their children; regrets about life; pain from a bad family relationship; pain from strong pride; pain from fluctuating decision making about medical treatment for respiratory problems; and pain from not being able to express gratitude to others. It is important for nurses to apply the notion of total pain to patients with ALS receiving NPPV

and to share their pain from early stage to the end with hospital and community care staff.

## ACKNOWLEDGEMENTS

The authors are grateful to the participant and the families of the deceased ALS patients. This work was supported by JSPS KAKENHI Grant Number 15K11617.

### *Declarations of conflicting interest:*

The authors declared no conflict of interest.

## REFERENCES

- [1]. Torres AL. The management of respiratory insufficiency in patients with ALS at or near the end of life. *Home Healthc Nurse* 2012;30(3):186-194.
- [2]. Societes Neurologica Japonica, ed.: Practical guidelines for amyotrophic lateral sclerosis. Nankodo Co., Ltd, Tokyo, 2013; 123.
- [3]. Tagami M, Kimura F, Nakajima H, et al. Tracheostomy and invasive ventilation in Japanese ALS patients: decision-making and survival analysis: 1990-2010. *J Neurol Sci* 2014;344(1-2):158-164.
- [4]. Kurisaki R, Yamashita S, Sakamoto T, et al. Decision making of amyotrophic lateral sclerosis patients on noninvasive ventilation to receive tracheostomy positive pressure ventilation. *Clin Neurol Neurosurg* 2014;125:28-31.
- [5]. Kimura F. Tracheostomy and invasive mechanical ventilation in amyotrophic lateral sclerosis: decision-making factors and survival. *Rinsho Shinkeigaku (Clin Neurol)* 2016;56(4):241-247 (in Japanese, Abstract in English).
- [6]. Quill CM, Quill TM. Palliative use of noninvasive ventilation: navigating murky waters. *J Palliat Med* 2014;17(6):657-661.
- [7]. Ushikubo M, Iida M, Suzuki M, Sasaki K. Literature review of palliative end-of-life care for patients with amyotrophic lateral sclerosis under non-invasive positive-pressure ventilation *KMJ*, 2017;66(4):55-62. (in Japanese, Abstract in English)
- [8]. Pizzimenti A, Aragona M, Onesti E. Depression, pain and quality of life in patients with amyotrophic lateral sclerosis: a cross-sectional study. *Funct Neurol*. 2013; 28(2):115-9.
- [9]. Xu J, Nolan MT, Heinze K, et al. Symptom frequency, severity and quality of life among persons with three disease trajectories: cancer, ALS, and CHF. *Appl Nurs Res*. 2015; 28(4):311-5.
- [10]. Pagnini F, Lunetta C, Banfi P, et al. Pain in amyotrophic lateral sclerosis: a psychological perspective. *Neurol Sci*. 2012; 33(5):1193-6.
- [11]. Chió A, Mora G, Lauria G. Pain in amyotrophic lateral sclerosis. *Lancet Neurol*. 2017; 16:144-157.
- [12]. Handy Cr, Krudy C, Boullis N, et al. Pain in amyotrophic lateral sclerosis: a neglected aspect of disease. *Neurol Res Int*. 2011; doi:10.1155/2011/403808
- [13]. Chió A, Canosa A, Gallo S, et al. Pain in amyotrophic lateral sclerosis: a population-based controlled study. *Eur J Neurol* 2012
- [14]. Ganzini L, Johnston WS, Shilveria L. The final month of life in patients with ALS. *Neurology* 2002;59(39):428-431.
- [15]. Mehta A, Chan L. Understanding of the concept of "Total Pain": A Prerequisite for Pain Control. *J Hosp Palliat Nurs* 2008;10(1):26-32.
- [16]. Saunders CM. The management of terminal malignant disease (2<sup>nd</sup>). London: Edward Arnold. 1984; 232-241
- [17]. Ushikubo M, Tomita C, Inokuma A, et al. Illness course and circumstances of death among individuals with rapidly progressive amyotrophic lateral sclerosis, *International Medical Journal*. 20(4), 2013:446-449.
- [18]. Cordess V, Sidorok F, Schimmel P, et al. Coordinated care affects hospitalization and prognosis in amyotrophic lateral sclerosis: a study. *BMC Health Serv Res* 2015; Doi:10.1186/s12913-015-0810-7.