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Palliative Care in Japan for Individuals with Amyotrophic Lateral Sclerosis

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Abstract

Palliative care has not been focused on noncancer yet. Amyotrophic lateral sclerosis (ALS) is a fatal, rapid progressive, and intractable neurodegenerative disease. Individuals with advanced ALS cannot perform activities of daily living by themselves, but their mental awareness remains clear. Individuals with ALS experience various pain soon after diagnosis, and a multidisciplinary team approach is needed because their pain can become complicated. Caring for individuals with ALS needs to both start and end with palliative care in the physical, psychological, social, and spiritual aspects. The Japanese Ministry of Health, Labour and Welfare enacted the Principles of Policy for Rare and Intractable Diseases (*nanbyo*), which were the first set of such principles to be established in the world. This chapter describes the palliative care in Japan, from the perspectives of nursing and *nanbyo* measures. Also, three unique characteristics of ALS care in Japan are mentioned: the high level of disagreement between patients and family; the high rate of ventilator use compared with Western countries; and the low consumption of morphine. Healthcare practitioners need to apply the notion of the total pain to provide palliative care to individuals with ALS and acknowledge the challenges of providing timely symptomatic management.

Keywords: ALS, palliative care, total pain, end-of-life care, family care, home care support system

1. Introduction

Amyotrophic lateral sclerosis is the most common degenerative disease of the motor neuron system. This disease is also known as Lou Gehrig's disease. The symptoms and progression processes can be quite varied in different people.

1.1. Characteristics of amyotrophic lateral sclerosis

Amyotrophic lateral sclerosis (ALS) is a devastating, neurodegenerative, and progressive disease with no established treatment. ALS is characterized by muscle weakness and atrophy, and these affect the upper and lower limb movements, swallowing, speech, and respiratory function. However, bowel, bladder, and oculomotor function and sensation are spared. The cognitive and behavioral symptoms have also been described, although frank dementia is a rare occurrence [1]. Patients with ALS may die within 3–5 years after the onset of their first symptoms if they do not choose to live with a ventilator [1]. Bulbar onset type, increased age, and front temporal dementia (FTD) are prognostic factors for shorter survival [2]. The course of ALS is quite uncertain, and sudden death or unexpected death is often seen [3]. Therefore, healthcare providers should consider end-of-life care soon after the diagnosis [3].

1.2. Epidemiology of ALS in Japan

The number of those patients with ALS has increased remarkably. **Figure 1** shows the number of patients with ALS who were certified as eligible for receiving medical financial aid in Japan for the treatment of a designated rare and intractable disease, which increased from 860 individuals in 1979 to 9950 individuals in 2015 [4].

The incidence of ALS in the Japanese population is much lower than in the Caucasian populations of Europe and North America. The prevalence and incidence rates per 100,000 people

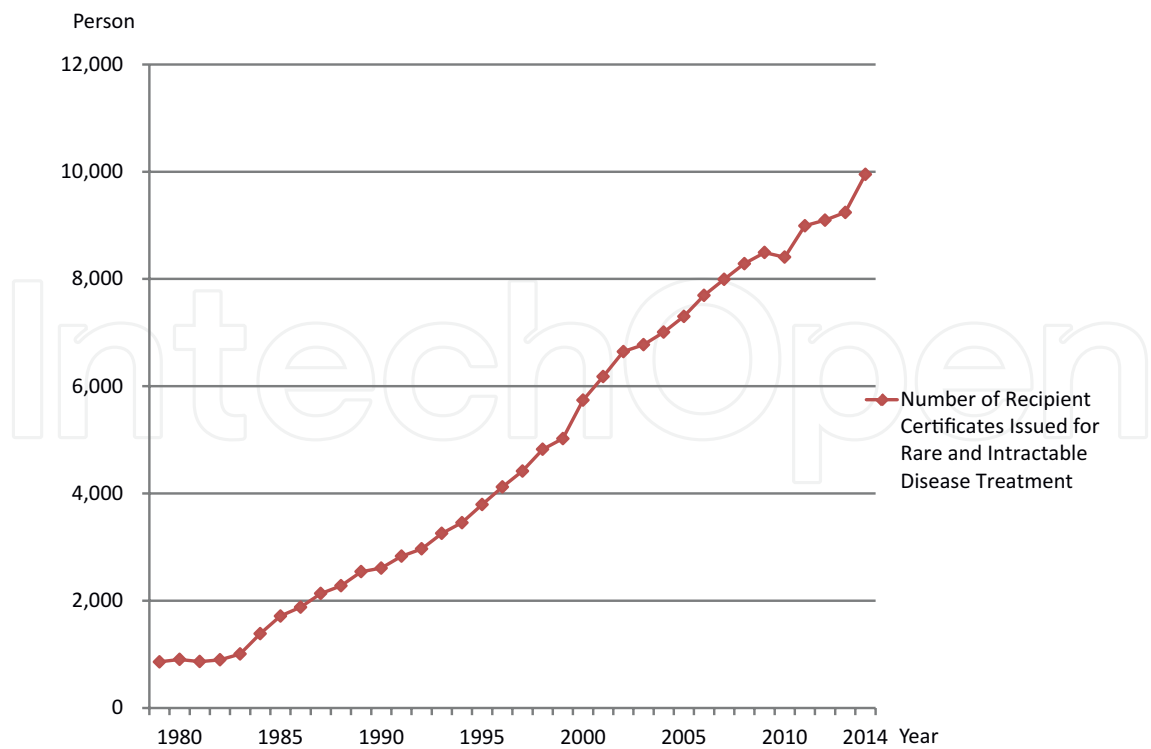


Figure 1. The number of ALS patients who are issued certificates in Japan for the treatment of rare and intractable disease between 1979 and 2015. Data are cited from Japan Intractable Disease Information Center [4].

per year in Japan were 9.9 and 2.2, respectively, in 2013 [5]. The highest prevalence, as well as incidence, was evident in the group comprised of individuals in their 70s, and the ratio of men to women was approximately 1.5 [5].

The aspects of ALS have also changed. A single-facility retrospective study conducted in rural Japan reported the following findings [6]. The percentage of ALS patients whose onset type was bulbar increased from 14.2% (1978–1982) to 38.3% (2008–2012) over the 35-year period of the study. The percentage of ALS patients with dementia increased from 0% (1978–1982) to 20.2% (2008–2012). The percentage of patients whose age at onset was ≥ 70 years increased from 0% (1978–1982) to 38.2% (2008–2012).

2. Continuous loss and decision-making accompanied by the disease progression of the disease characteristic in individuals with ALS

Individuals with ALS suffer from decreasing function in their activities of daily living (ADL), such as immobility, respiratory insufficiency, speech difficulties, and swallowing difficulties. Poor ADL performance leads to care burden. Physical pain in individuals with ALS is not proportional to the severity of the disorder [7]. Psychological distress is great even in the early stage shortly after diagnosis [8]. The reactions of the patients and their family members at the time of diagnosis need to be shared with the hospital and community care staff in order to start providing adequate support. Individuals with ALS can develop a sense of security and suppress cumulative suffering if they feel that they are supported from an early stage.

Individuals with ALS cope with relentless loss in the disease progression stage. When disease progression is fast, the patient often loses additional bodily functions before his or her illness acceptance can catch up. The patient's psychological state can be unstable due to the steady loss in physical function at a time when various difficult decisions (e.g., when and how to utilize social services and undergo medical treatment) must be made by the patient and his or her family [9].

2.1. Palliative care for ALS symptoms and pain

The symptomatic management should be provided in a timely manner in order to slow progression and optimize quality of life (QOL). It can be said that care for individuals with ALS starts and ends with palliative care. Symptoms of ALS which need palliative care include motor dysfunction, dyspnea, dysphagia and drooling, dysarthria, and pain. Other symptoms such as depression, anxiety, sleep disturbances, and nightmares must be addressed with specific pharmacological and non-pharmacological approaches including a supportive psychotherapy, which is usually best administered as family therapy.

2.1.1. Motor dysfunction and palliative care

It causes mental distress that patients cannot perform ADL by themselves and need to be cared for by others. Individuals with ALS experience repeated falls as physical function deteriorates,

which forces wheelchair dependence and eventually progresses to a bedridden condition. Living with this deterioration and disturbance in one's body image is also painful. These patients lose their jobs, and the opportunities for going out and socializing are inevitably reduced. The range of life naturally becomes narrower. Accordingly, motor dysfunction develops into a spiritual pain whereby individuals with ALS question the meaning of life, thereby exacerbating psychological and social distress [10].

Healthcare practitioners need to respect and focus on the residual abilities of the individuals with ALS—not the parts that they cannot do—and make the utmost use of what they can accomplish. It is important to find and devise a way of life with the individual with ALS and his or her family members that incorporates adequate assistive devices or utensils so that tasks can be performed by the individuals with ALS.

Although the people surrounding individuals with ALS tend to thoughtlessly reach out because of feelings of pity and kindness and prevention from danger situations, these actions can increase their mental distress of the individuals with ALS and reduce their self-esteem [10]. Technology development makes individuals with ALS do themselves by operating environmental devices, as long as they have a part of the body that they can move slightly, such as the tip of their little finger or eye tracking.

Human value is not changed at all even if the body cannot completely move. It is important for individuals with ALS to have roles and feel that he or she is needed from their surrounding friends and family. The healthcare profession needs to build up partnerships with their patients and their family members in order to enhance their autonomy and power to live.

In addition, active and passive physiotherapy has a pivotal role, especially in order to prevent muscle contractures and joint stiffness, whereas acetylcholinesterase inhibitors (e.g., pyridostigmine) may lead to a short-term improvement in muscle strength, especially during the early stages of the disease and in those with bulbar symptoms.

2.1.2. Respiratory dysfunction and palliative care

If breathing cannot be adequately performed, this will directly lead to a life crisis. Patients express the respiratory disorder caused by ALS as “feeling [like I am] drowning in the sea while in the land,” “feeling [like I am] being strangled with cotton,” and “suffering [because I] cannot cough up phlegm.” The leading cause of death of individuals with ALS is respiratory failure, and they must live with the fear of approaching death all the time.

For the early detection of respiratory disorders in homecare, healthcare practitioners need to perform peripheral oxygen saturation (SpO₂) checkups during the nighttime and measure sniff nasal inspiratory pressure, the difference between chest girth at expiration and inspiration, and forced vital capacity. Symptomatic treatments such as respiratory rehabilitation, mechanical insufflation-exsufflation (MI-E), noninvasive positive pressure ventilation (NPPV), tracheostomy, and tracheostomy positive pressure ventilation (TPPV) can also be applied.

Ideally, NPPV should be introduced before the respiratory muscles are excessively exhausted. However, some patients do not undergo the early introduction of NPPV because they believe

that they “do not want to live by relying on any machine” or “if [they] rely on machines, [their bodies will get] stuck.” In addition, sometimes the NPPV therapy itself is only troublesome when they are not well aware of respiratory insufficiency and cannot realize the effect of NPPV.

Healthcare practitioners need to explain how symptoms can be relieved, rather than just prolong life, in order to promote a better adaptation and tolerance to NPPV. Individuals with ALS are not able to detach and attach the NPPV mask whenever they desire due to upper limb disorders. Since displacement of the mask can cause death, [3], a sufficient home care support system must be prepared for each NPPV user.

Also, a small amount of morphine provides effective relief for respiratory distress in patients with ALS [11]. Morphine consumption in Japan is lower than in other countries. According to 2008–2010 data compiled by the Ministry of Health, Labour and Welfare, morphine consumption was 204.5 g/day per 1 million persons in the United States, 191.8 g/day per 1 million persons in Canada, and only 7.2 g/day per 1 million persons in Japan [12]. The use of morphine for neurological diseases was approved by the Japanese national health insurance system in 2011 [13]. Morphine consumption can be expected to increase in the near future.

2.1.3. Difficulty in swallowing and eating and palliative care

Oral intake can become difficult as ALS progresses. Poor oral intake or dysphagia as well as difficulty with self-feeding and meal preparation leads to weight loss and malnutrition and increases the risks of aspiration pneumonia and suffocation. Moreover, poor oral intake deprives the pleasure of individuals with ALS and lowers QOL. The long time required for oral ingestion leads to fatigue. Malnutrition causes not only respiratory waste but also the loss of spirit need to think or redesign the way of future life [14].

There are many ways to increase oral intake: consider the position of the neck and head, change the patient’s body posture, modify the type of food, use a thickener, conduct swallowing rehabilitation therapies, perform an esophageal laryngectomy or total laryngectomy, etc. It is also important to collaborate with professionals such as speech therapists, dentists, and certified nurses who specialize in dysphagia. Percutaneous endoscopic gastrostomy placement is also indicated.

2.1.4. Difficulty in speech and palliative care

Due to the disease progression, individuals with ALS are difficult to say what they want to say. The individuals with ALS frustrate that the things they want to convey do not pass well and experiences a sad feeling by being told that “I do not know what you are talking about” from others. Individuals with ALS may lose their voice by tracheostomy.

Communication greatly affects QOL of the individuals and their family. In recent years, remarkable progress has been made in the field of augmentative and alternative communication (AAC), which includes all forms of communication (other than oral speech) that are used to express thoughts, needs, wants, and ideas. Consequently, various communication devices

and switches are available. In cooperation with speech therapists and occupational therapists, selection and contrivance of a communication device is required along with aggravation of the disease condition.

As training and time are necessary to use communication devices, it is desirable to introduce them at an early stage where the disease state is mild, but until patients do not realize the necessity, they are reluctant to introduce. It is an important key whether or not the individuals with ALS desire to convey their thoughts and whether or not their family and supporters feel like wanting to hear the individual’s thoughts.

2.1.5. *Physical pain*

The two types of physical pain are summarized in **Table 1**. Although fasciculation—arising through degeneration of the intramuscular motor axons—and subsequent painful muscle cramps are often the first symptoms of the disease, the secondary in nature is most reported types of pain [15].

2.1.6. *Total pain*

As mentioned above, deterioration of physical function and various types of physical pain cause mental, social, and spiritual distress. These pains become increasingly complicated and cumulatively expand with disease progression. Therefore, the concept of “total pain” coined by

Primary forms of pain
<ul style="list-style-type: none">• Neuropathic pain (extremities)• Painful cramps (lower limbs, hands, abdomen)• Spasticity (lower limbs)• Pain caused by inability to move and change position (diffuse, including buttocks, limbs, and trunk)• Paresis of limbs (shoulder pain, articular pain)• Joint contractures (hand and ankle joints)• Itch (diffuse)
Secondary nature (most reported types of pain)
<ul style="list-style-type: none">• Neck pain due to hyposthenia of neck muscles• Back pain due to reduced mobility• Pressure sores due to reduced mobility• Pain due to suctioning of phlegm• Articular pain due to reduced mobility• Shoulder pain due to reduced mobility• Facial pressure ulcers due to NIV mask• Pain due to ventilator hose pulling or weighing

Table 1. Types of pain in individuals with ALS, adapted from Chió et al. [15].

Dame Cicely Saunders to manage the cancer pain—which has physical, psychological, social, and spiritual components whereas the contribution of each component will be specific to each individual and his or her situation [16]—can also be applied to rare and intractable diseases [9, 13]. Pain will never be relieved by only paying attention to physical aspect (**Figure 2**). That is why individuals with ALS should be supported using a multidisciplinary team approach.

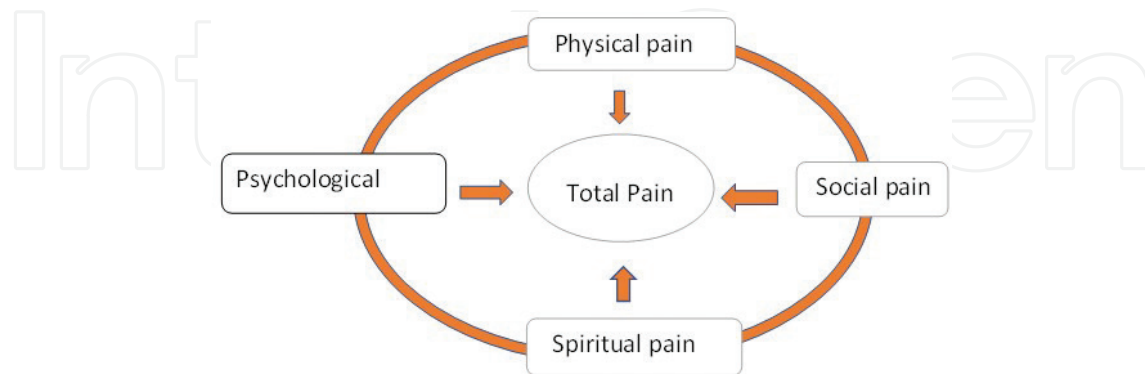


Figure 2. Total pain, adapted from Ushikubo [9].

3. Government measurements in Japan

3.1. Definition of “Nanbyo”

Rare and intractable diseases are called *nanbyo* in Japan. *Nanbyo* is defined by the following four conditions: (1) no known cause, (2) no clearly established treatment, (3) rare disease status, and (4) long-term care is typically required [4]. *Nanbyo* is the administrative term that the government uses to promote clinical investigations and research in order to establish effective treatments and improve cares [4]. Patients are often shocked by the poor image of the term *nanbyo*. However, they should understand the term in a positive sense. *Nanbyo* is the administrative term which has the meaning that the Japanese national government strengthens to clarify the causes of disease and effective treatments.

Patients registered as having one of the designated *Nanbyo* or the designated rare and intractable diseases are eligible to participate in a subsidy scheme that covers medical expenses. The designated *Nanbyo* selected from rare and intractable diseases that meet the following two conditions: (1) diseases with a prevalence of less than 0.1% of the population in Japan (i.e., affecting less than approximately 140,000 persons) and (2) diseases with established objective diagnostic criteria. As of April 2017, 330 rare and intractable diseases were designated [4]. The typical neuromuscular *nanbyo* includes ALS, multisystem atrophy, Parkinson’s disease, spinocerebellar disease, and multiple sclerosis.

3.2. Historical development of Japan’s governmental policy

The system of rare and intractable diseases in Japan was started ahead of the world as “*nanbyo* measures” and gradually getting improved [17]. In 1972, the Ministry of Health,

Labour and Welfare enacted the Principles of Policy for Rare and Intractable Diseases [4] which was the first set of principles to be enacted in the world. Since then, the government has promoted research and expanded support for individuals with rare and intractable diseases. At first, there were three pillars that were used to clarify causes and treatments: promoting surveys and research; establishing medical institutions; and reducing copayments for medical fees.

In 1995, the Principles of Policy for Rare and Intractable Diseases was revised to include an additional two pillars: improving cooperation between community-based healthcare, medical, and welfare agencies; and promoting welfare measures aimed at improving QOL [4].

The characteristic of *nanbyo* is disease progression, while disability is defined as fixed disorders. Individuals with *nanbyo* had not been recognized as the disabled based on the Act of Services and Supports for Persons with Disabilities. Since this Act was amended in 2013, individuals with *nanbyo* patients have been recognized as the disabled [18]. As a result, the use of disability welfare services (e.g., support consultations, prosthetic devices, and community life support projects) has been expanded to individuals with *nanbyo*.

According to the increasing number of people with intractable diseases, the budget required for medical expense subsidies has also expanded. In addition, from the viewpoint of fairness, there was a demand for the further expansion and review of medical expense subsidies for people with other diseases which had not been defined as the designated *nanbyo*. More than 40 years have elapsed since the establishment of the Principles of Policy for Intractable Disease, and the “law concerning medicine etc. of intractable patients (*nanbyo* law)” [19] has been enforced since January 2015. This *nanbyo* law made stabilized medical expense subsidies by allocating financial resources such as a consumption tax [18, 19]. In addition, the designated *nanbyo* (i.e., the targeted diseases eligible for medical expenses subsidies) has been expanded from 56 diseases to 330 [17].

3.3. Social services

Public social services—which *nanbyo* patients can utilize—are gradually increasing. Individuals with *nanbyo* are available to utilize public social services by combining the following four systems: medical insurance, long-term care insurance, the welfare system for persons with disabilities, and the *nanbyo* medical system. Since the different offices deal with each system, it is not easy to use them. However, these four systems are very proud of in the world by the fulfilling contents [19].

Even if healthcare practitioners encourage individuals with *nanbyo* to use social resources, they may be reluctant to initially propose the use of social resources. Multiple factors are implicated in the uptake of social services [20]. ‘Internal’ issues focused on retaining control and normality within the home. Suggesting an individual use social services can be perceived as insulting because it indicates that the individual cannot provide his or her own medical care good enough or family care is insufficient. Healthcare practitioners must consider the patient’s and family’s feelings whenever suggesting the use of social services and should research reasonable methods for proposing the use of services.

4. Home care support system for individuals with ALS

Figure 3 shows the system for providing home care support during the early stage following diagnosis; **Figure 4** shows the system used during the late and severe stages. The types and frequencies of use for various social services should be considered along with both disease aggravation and the preferences of the individual with ALS and his or her family.

Collaboration among multidisciplinary professionals is extremely important. Because nurses specialize in both medicine and care, they play the role of the glue that holds together the multidisciplinary team [21].

The concept of hope is important to supporting individuals with ALS. Healthcare practitioners need to seek and sustain their hope in individuals with ALS. Individuals with ALS have hope for a cure, social support, searching for information, spiritual beliefs, limiting the impact of the disease, adapting to changing abilities, and self-transcendence [22].

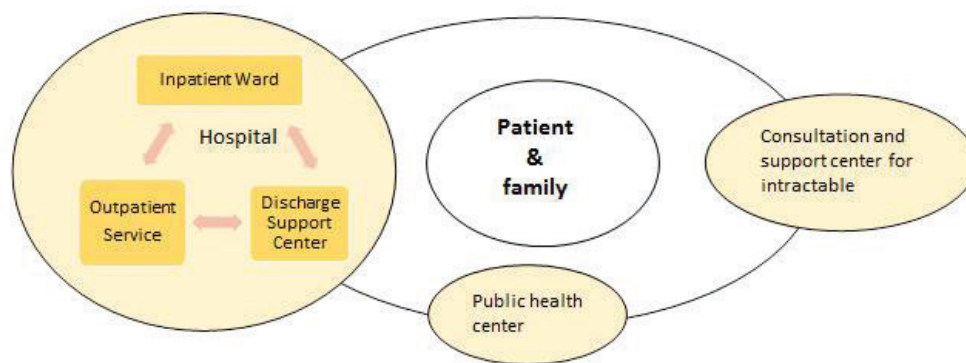


Figure 3. Support system for individuals with early stage ALS.

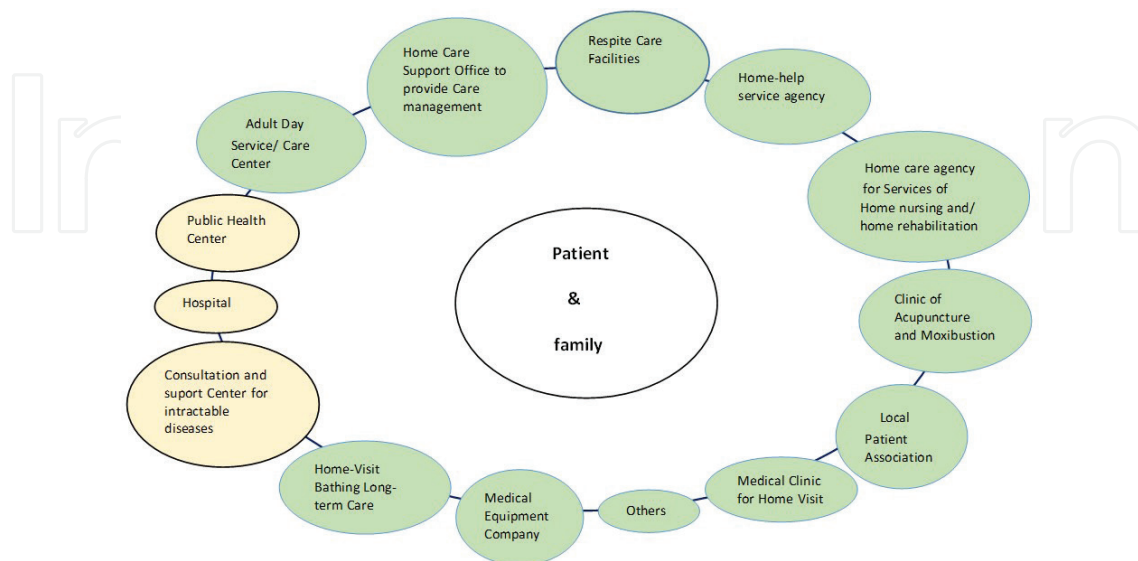


Figure 4. Social services and home care support system for the individuals with ALS during disease progression.

5. Timely decision-making for medical treatment

Although there is no effective cure for ALS, symptomatic treatment can improve QOL if it is provided in a timely manner. Also, the healthcare provider can modify the illness trajectory, hinder disease progression, and extend survival time. Providing symptomatic treatment is essential in order to gain the patient's understanding and consent. Healthcare practitioners need to understand the reasons why psychological distress of individuals with ALS is generated by social interaction.

Individuals with ALS can hardly accept their diagnosis. As ALS rapidly progresses, their illness acceptance cannot catch up with the reality of disease condition. It is no wonder that patients expect to return their previous health and become well someday, while healthcare practitioners play the percentages based on their knowledge of the disease characteristics. Individuals with ALS suffer from this gap between them and their healthcare practitioners. Also, their families have the poor understanding of the reality of disease progression. This also allows increasing the suffering of the individuals with ALS [23].

Providing information is an important part of decision-making support during disease progression. Healthcare providers consider timing to be a key point in the decision-making process and often postpone decisions until it is not too late.

6. End-of-life care in Japan

6.1. Two terminal points in individuals with ALS

Individuals with ALS have two terminal points. As respiratory failure worsens, if the patient decides not to receive TPPV, then he or she will die at this primary terminal point. If an individual with ALS selects to receive TPPV at this primary terminal point, he or she will encounter the secondary terminal point at time that TPPV can no longer provide respiratory management.

Ten-year survival rate was 87.1% among individuals with ALS who underwent TPPV from 2000 to 2007 [24]. TPPV allows long-term survival for individuals with ALS, but patients with terminal cancer are limited to live longer even if TPPV is initiated. The other difference between cancer and ALS, as shown in **Figure 5**, is that ALS is living with fear of dying all the time, while cancer patient can change the gear from living positive to death.

6.2. Rates of TPPV use

The rate of use of TPPV is higher in Japan than Western countries, as shown in **Table 2**. Some reasons for this discrepancy are summarized in **Table 3**.

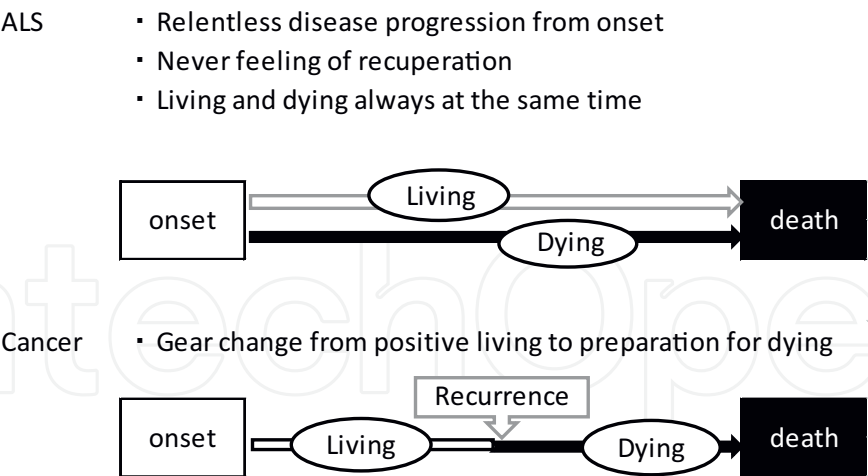


Figure 5. Difference between ALS and cancer.

Citation, which data were requoted	Country	Rate (%)	Study method
Furukawa et al. [25]	Japan	29.30%	National study
	Western countries	1.5–3.2%	
Tagami et al. [26]	Japan and Asian countries	12.7–21%	A British series of 50 patients
	Britain	0%	
	Germany	3%	
	USA	1.4–14%	
	Canada	1.50%	
	France	2.5–5%	
	Norway	7% for males and 3.8% for females	
	Italy	10.60%	
	Denmark	22%	
Rabkin et al. [27]	USA	2–6%	The largest study in north Italy from 1995 to 2004
	Europe	0–10.6%	
	Japan	25–46%	
Lee et al. [28]	Taiwan	20.97%	

Table 2. International comparison of TPPV rates.

Citation	Reasons
Tagami et al. [26]	<ol style="list-style-type: none"> 1. Attributed to religious, ethnic, and cultural differences 2. Financial considerations: the Japanese government covers all costs for noninvasive/invasive mechanical ventilation at any age for both inpatients and outpatients 3. Regular follow-up examinations at the same institute due to increased availability of continue services and improved knowledge and education regarding ALS 4. Peer counseling by the Japanese ALS Association (JALSA) 5. Implementation of the long-term care insurance (LTCI) as a new and fundamentally reformed social insurance system at 2000 in Japan
Rabkin et al.[27]	Different conceptualizations of the physician's role in medical decision-making, attitudinal differences among patients, caregivers and healthcare professionals and
Vianello et al. [29]	<ol style="list-style-type: none"> 1. To underline a proactive approach by Japanese physicians toward TV 2. Traditional fiduciary relationship that exists between the patient and the physician (the traditional Japanese paternalistic medical ethos dictating that "the doctor knows best" associated with physician's belief that patient well-being is the most) 3. Be attributable to the contribution offered by family members, given that the family's obligations as home care providers during sickness are deeply embedded in

Table 3. Reasons why Japan has a higher rate of TPPV use than other countries.

6.3. High level of disagreement between individuals with ALS and their family members in Japan

Differing opinions about the use of TPPV are often seen between individuals with ALS and their family members. Fifty-four percent of Japanese patients oppose TPPV compared with 10% of Japanese caregivers, but American patients and caregivers show greater agreement in decision-making than their Japanese counterparts [30].

There is no correct answer regarding decision-making. The most important thing on decision-making is what the individual with ALS thinks. However, Japanese individuals with ALS consider the burden of care on their family members to be greater than the burden on themselves and often do not want to receive TPPV. Families want individuals with ALS to receive TPPV because they want their loved ones to live as long as possible. This relationship, which considers each other, results in a large amount of disagreement between the individuals with ALS and their family members.

Having sufficient time to think and discuss will lead to the best decision-making. Healthcare professionals need to promote individuals with ALS discussing decision-making with the relevant family members [31]. Early intervention is necessary before the speech difficulties

associated with ALS become severe [30]. A study on American and Canadian patients with ALS reported that advanced directives were in place for 88.9% of patients and followed for 96.8% of patients [32]. However, advanced directives have not been familiarized yet in Japan [33].

Reference	Published Year	Country	Method	Place of death
Gil et al. [35]	2008	France	<ul style="list-style-type: none"> • 302 patients in one of the 16 ALS centers in 15 French regions • Prospective investigation 	63% of patients died in a medical facility
Yang et al. [36]	2011	China	<ul style="list-style-type: none"> • 139 patients in West China Hospital of Sichuan Univ. • Survey to family members, caregivers, family physicians 	82% of patients died at home
Spataro et al. [37]	2010	Italy	<ul style="list-style-type: none"> • 182 patients in a single ALS tertiary center • Consultation with relatives or family physicians 	85.2% of patients died at home
Ushikubo [38]	2015	Japan	<ul style="list-style-type: none"> • 14 patients in users of home care nursing • Interview survey with home care nurses 	43% of patients died at home
Dominguez-Berjon et al. [39]	2015	Spain	<ul style="list-style-type: none"> • 1035 in the Autonomous Community of Madrid • A population-based cross-sectional study 	56.1% died in a hospital 30.4% at home, 8.3% in a residential home 5.1% unknown
Escarrabill et al. [41]	2014	Portuguese	<ul style="list-style-type: none"> • 77 from five hospitals • A retrospective medical records analysis 	57% of patients died in a home
Tsai et al. [42]	2013	Taiwan	<ul style="list-style-type: none"> • 751 from the National Health Insurance Research Database • A retrospective population-based study 	53.5% died in a medical facility

Table 4. International comparison of the places of death of individuals with ALS.

6.4. End-of-life care for patients with ALS receiving NPPV

It is true that palliative end-of-life care for individuals with ALS has not progressed much in Japan [33, 34]. People who use NPPV are increasing because it is a way to palliate respiratory deficiency and give time for decision-making on TPPV. Euthanasia is not legally permitted in Japan. However, NPPV cannot palliate respiratory failure if the disease is severely aggravated [35]. Palliative end-of-life care for patients who choose to remain on NPPV as the ceiling of medical treatment for respiratory failure has received little focus. The development of palliative end-of-life care for ALS patients receiving NPPV is needed [35].

6.5. Place of death among individuals with ALS

Understanding the causes and places of death of ALS patients is necessary to develop better end-of-life care. There are seven articles [36–42] that clarify the causes and places of death. The primary cause of death remains respiratory failure. The secondary causes of death vary and include sudden death, death during sleep [38], nutritional causes [37], and cardiovascular diseases [42]. **Table 4** summarizes seven articles on the places of death of individuals with ALS, and these also vary. Age, marital status, and the socioeconomic level of the patient also influence the places of death of patients with ALS [40]. Further research is necessary to develop end-of-life care by elucidating the dying process and analyzing the place of death.

7. Conclusion

There is no effective cure for ALS, but symptomatic treatment accomplishes the purpose of palliative care, extends the patient's survival time, and improves their QOL. Healthcare practitioners have many ways to improve illness trajectory and QOL by coping with the various symptoms caused by disease progression in a timely manner. It is important to provide mental care, decision-making support, and family support via multidisciplinary collaboration. Healthcare practitioners make an effort to cherish hope and support the autonomy of the individuals with ALS and their family members. Moreover, it challenges to develop and provide palliative care on the terminal withdrawal of NPPV.

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