Reexamining the capabilities of ALS patients

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Abstract

This paper discusses the capabilities of patients with Amyotrophic lateral sclerosis (ALS). First, we describe the general circumstances of ALS patients in developed countries, and give a brief explanation of Japan's medical and long-term care system that partly succeeds in broadening the capabilities of ALS patients. Second, we examine clinical research studies about ALS. Third, we introduce three studies we have conducted that focus on the improvement of the capabilities of ALS patients on ventilators. Our conclusion is that medical and social support systems should not ignore the fact that with sufficient care some advanced ALS patients can enjoy a reasonably high quality of life without placing an undo burden on their families.

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1. Introduction

Well-being and agency

Sen(1985)(1987)(1992) has broadened the informational basis of the idea of freedom utilizing the distinction between well-being freedom and agency freedom. According to Sen, well-being freedom is freedom to access the bundle of functions that consist of people's well-beings. Agency freedom, on the other hand, is "freedom to achieve whatever the person, as responsible agent, decides he or she should achieve[6]", and "that open conditionality makes the nature of agency freedom quite different from that of well-being freedom, which concentrates on a particular type of objective and judges opportunities correspondingly[7]".

This distinction between the well-being and agency aspects of human freedom gives us clearer insight about the idea of not only freedom but also happiness, justice and equality of human beings and has greatly influenced studies related to the capability approach.

One feature of agency freedom is that people sometimes sacrifice their own well-being to realize their objectives as agents. Capability theorists including Sen often refer to such situations, citing examples such as fasting for the sake of political protest. However, at the same time Sen insists that respecting a person's agency freedom doesn't justify ignoring her well-being, even though she may herself in some cases do so .

> The fact that you may have chosen the 'perfection of the work' over that of your 'life', or have chosen to give priority to other goals in your agency objectives over your own well-being is not a reason to think that your life or your well-being is henceforth of no importance—to you or to others[8].

> A self-sacrificing idealist who is ready to sacrifice fully his own well-being for some 'cause' does not thereby make it okay for others to ignore his well-being so long as his 'cause' is not harmed[9].

ALS patients' "Life or death" decision

Recently numerous studies have been reported concerning the "freedom to die", "desire to

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[6] Sen(1985:203-204)
[7] Sen(1985:203-204)
[8] Sen(1992:70)
[9] Sen(1992:71)

die" and "care before death" of patients with Amyotrophic Lateral Sclerosis, ALS[10]. Many clinical research studies have focused on investigating personal and social characteristics of patients and their families, their difficulties related to ALS, and their attitudes toward physician-assisted suicide(PAS), euthanasia, and receiving ventilators. Some of them then discuss what implications their data have for the question of whether PAS, euthanasia and ventilators should or should not be employed.

It is potentially misleading, however, to discuss ALS patients' clinical issues without considering the possibility of improving the well-being of ALS patients through effective social support. While it is important to examine the agency aspect of ALS patients' decisions, in particular their preference in regard to dying or using mechanical ventilation to prolong their lives, to do so without considering the full extent to which their well-being freedom can be improved, in other words, to assume that the disease itself inevitably leads to an low quality of life, is to ignore an important part of the picture.

To consider this problem, our discussion has been divided into three parts. First, we describe the general circumstances of ALS patients in developed countries, and give a brief explanation of Japan's medical and long-term care system that partly succeeds in broadening the capabilities of ALS patients. Second, we examine clinical research studies about ALS. Third, we introduce three studies we have conducted that focus on the improvement of the capabilities of ALS patients on ventilators.

2. ALS in Japan

What is ALS?

According to the J*ournal of the American Medical Association, JAMA,*

> Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig disease, involves progressive loss of motor neurons (a type of nerve cell controlling muscle movements) in the brain and spinal cord. ALS is a progressive, disabling, and ultimately fatal disease of unknown cause. Walking, speaking, swallowing,

[10] ALS is a "disease that causes gradual paralysis and respiratory failure and that result in death within three to five years, on average, after the diagnosis has been established." (Ganzini, et al. (1998: 967)). We will quote a more detailed explanation in section 2 and also point out some features of ALS that are not mentioned usually.

> breathing, and other basic functions become impaired with time. About 30 000 Americans currently have ALS. The yearly incidence rate is 1 to 2 new cases per 100 000 individuals. The disease is commonly discovered during middle age and affects more men than women[11].

Therefore, ALS patients die of malnutrition or respiratory failure if feeding tubes[12] and ventilators are not used at a certain stage. Because patients' bodies completely lose their ability to perform certain functions necessary for survival, it is normally said that "death occurs within 3 to 6 years after symptoms begin[13]" or that ALS "results in death within three to five years, on average, after the diagnosis has been established[14]". However, if patients decide to receive feeding tubes and a ventilator, they are in fact often able to live much longer.

ALS treatment in developed countries

In developed countries, the majority of ALS patients don't proceed to Long-Term Mechanical Ventilation(LTMV) or receive LTMV just temporarily. This means that they choose to die rather than to go on living with the aid of ventilators[15]. Therefore, ALS is generally considered a "terminal" disease that inevitably leads the death of those who suffer from it.

One reason that many ALS patients decline LTMV is that most doctors, patients, and families assume that living with advanced ALS and LTMV for a long time is not a realistic option because the QOL(Quality of Life) and well-being of both the patients themselves and their families becomes very low.

While this assumption is present in Japan, in the last thirty years living on LTMV with long-term home care has become more widely considered as a viable option, and in fact 30% of ALS patients in Japan are receiving LTMV treatment. This rate is significantly higher than the rate in western countries and it appears to be increasing[16]. Mitsumoto et al.[2007]

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[11] http://jama.ama-assn.org/cgi/reprint/298/2/248 in JAMA Patient Page

[12] Feeding tube, formally called Gastorostomy, is a method to inject nutrition directly into a stomach using a penetrating tube.

[13] http://jama.ama-assn.org/cgi/reprint/298/2/248 in JAMA Patient Page

[14] Ganzini, et al. (1998: 967)

[15] Miller(2006), Oliver et al.(2006)

[16] Mitsumoto et al.(2007)

suggests that this noticeable difference is related to "financial considerations(the Japanese government covers costs of LTMV), physician support for LTMV, social pressure from the Japanese ALS Association, and differences in cultural attitudes about "truth-telling" and death"[17].

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Prevalence of LTMV treatment in Japan

The prevalence of LTMV treatment in Japan can be attributed to the "Outline of measures for Nanbyo" that was announced by the Japanese Ministry of Welfare in 1972. At that time the Japanese government had organized a research study committee to analyze the cause of a strange disease named SMON (Subacute Myelo-Optico-Neuropathy). After the committee elucidated the mechanism of SMON disease, Neurologists and other specialists in the committee continued studying other neurological diseases and worked together to improve the treatment and support of patients suffering from neurological disorders. As a result, the Japanese government officially defined some incurable diseases that seriously harmed patients' bodies and social lives as "Nanbyo[18]" and started allocating a budget on a consistent basis to promote research projects and reduce the medical costs born by patients suffering from "Nanbyo".

The number of "Nanbyo" recognized by the government has gradually increased, and ALS become one of them in 1974. Since then, high medical expenses previously born by patients have been covered by national and local governments[19] and medical treatment and home care has been promoted. Consequently, some ALS patients on LTMV have been able to realize relatively high QOL when sufficient care is available.

Conventional medical treatment of ALS regards respiratory and swallowing disorders as "terminal stage". But the governmental "Nanbyo" support program and the actions of ALS patients, families and specialists in Japan have partly created a paradigm shift that has changed the idea of "terminal stage" to "long-term treatment stage on LTMV".

This shift was in part fostered by several support groups for ALS patients that have been organized in Japan. The Japanese ALS Association, JALSA, the biggest support group, was

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[17] Mitsumoto et al.(2007)

[18] Directly translated "Nan-byo" means "difficult diseases" Recent requirements for a disease to be classified as a "Nanbyo" are rarity, being of inexplicable cause, incurability, and causing long-term difficulties in daily life.

[19] Though the insured in Japanese national health insurance normally has to pay 10 to 30% of medical costs as his or her own expenses, all the expenses of ALS patients are covered by national and local government.

founded in 1986. JALSA, which includes peer support groups run by ALS patients themselves, considers LTMV as a positive tool to lengthen patients' lives and improve their QOL and has applied pressure to the Japanese government in order to obtain the necessary social support for long-term treatment with LTMV. In other words, JALSA has taken quite a different approach from other countries' ALS associations that basically assume that living on LTMV inevitably leads to low QOL for patients and their families.

A new social care scheme for people with disabilities, called *Shienhi Seido*, was implemented in Japan in 2003[20] and ALS patients were accredited as persons with severe disabilities under the scheme. With these new form of support, ALS patients on LTMV have been able to employ more than one caregiver when they go out, and some patients in the Tokyo metropolitan area have been able to receive continuous 24-hour a day care.

In areas in which sufficient medical treatment and social care are available, a few ALS patients have been able to live totally independently from their families and 13 ALS patients and their families have started home care businesses. Their activities have been used as the basis for the "Sakura Model[21]", an ALS version of the "Independent Living Movement" paradigm[22]. One of the Sakura Model's outstanding features is that this program fully utilizes paid care givers and realizes not only patients' but also their families' well-being by releasing families from the burden of providing intensive care.

3. Previous clinical research studies

Looking at the history of ALS in Japan, it is clear that the perception of LTMV has been a critical factor that has influenced medical and socio-economic policy surrounding this disease and its treatment.

As we mentioned above, ALS is usually considered to be an incurable disease that results in death within several years[23]; respiratory failure is basically regarded as a fatal result of ALS and receiving LTMV is considered exceptional. Though some recent studies treat LTMV as a form of palliative care and a few studies have investigated the effects of LTMV on ALS patients, it is still difficult for most patients to decide to live on LTMV.

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[20] This scheme was changed to a similar but different system in 2006.
[21] The Sakura Model was created by members of Sakura, an NPO supporting ALS patients. "Sakura" means "cherry blossom" in Japanese. See Kawaguchi(2006b) for more details.
[22] For more on the "Independent Living Movement" see Barnes et al.(1999).
[23] Ganzini. et al.(1998: 967).

To illustrate this situation, we have classified previous studies on the topic into four groups. The first group consists of studies that mainly focus on the situations[24] and QOL[25] of ALS patients. The second group surveys patients' attitudes toward death, physician assisted suicide and euthanasia[26]. The third group consists of studies that focus on the well-being of ALS patients on LTMV and their families[27]. The fourth group contains studies that refer to LTMV or make LTMV a central topic, but mainly focus on informed consent and advanced directives with regard to whether a patient wants to receive LTMV or not[28].

In summary, the first and second groups basically do not focus on LTMV problems and some studies seem to assume that there is little potential for ALS patients to live with reasonably high well-being on LTMV. This underlying attitude can be perceived in comments like the following.

> "Having control over one's death may be especially important for persons with a disease such as ALS, in which the inability to work, engage in pleasurable activities, care for oneself, and communicate constitutes a formidable loss of autonomy."[29]

On the other hand, the third or forth groups look into ALS patients on LTMV more precisely and some studies in the third group show that most informants surveyed had positive attitudes toward LTMV. For example, Moss et al.(1993) found that "90% of patients were glad they had chosen home ventilation and would choose it again". Gelinas, et al. (1998) reported that "no patient regretted the decision to go on the ventilator and most patients felt contented and satisfied a majority of the time" and that patients "reported greater happiness when they could go out of the home and participate in other social activities." Kaub-Wittener, et al. (2003) pointed out that "the vast majority of ALS patients on NIV[30] refuse tracheostomy" but "81% of TV[31] patients would choose ventilation again."

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[24] For example, Krivickas et al.(1997) , Goldstein et al.(2000) ,Mandler et al.(2000), Rabkin et al.(2000)(2005) measures several psychiatric parameters or families' care burden.
[25] These studies includes Gelinas et al.(1998) , Jenkinson et al.(2000), Simmons et al.(2000), Robbins et al.(2001), Lyall et al.(2001), Bromberg and Forshew (2002), Lou et al.(2003), Bourck et al.(2003)
[26] Ganzini et al. (1998)(2002), Bascom & Tolle (2002),Veldink et al.(2002) and Albert et al.(2005)
[27] Borasio et al.(1998), Gelinas, et al.(1998), Kaub-Wittener, et al. (2003) and partly Chio et al.(2004)(2005)
[28] Moss, et al. (1996), Albert et al. (1999), Rabkin,et al.(2006), Mitsumoto et al.(2007)
[29] Ganzini, et al. (1998: 971). Veldink, et al. (2002: 1643) also refer to the same phrase.
[30] Non-invasive ventilation(NIV) via mask. is 'now recognized as a standard in ALS care'

4. Three studies examining patients' attitudes and experiences concerning LTMV

Background and objectives of studies

As mentioned above, some studies on LTMV show that ALS patients on LTMV can achieve reasonably high well-being and positive attitudes toward LTMV. But they do not make clear how it is possible for ALS patients on LTMV to achieve this well-being in spite of their severe physical disabilities. We have conducted several studies to explore this issue, and summarize the results of three of them below. The first two studies point out some factors that affect the well-being of ALS patients on LTMV, and the last study describes how sufficient care has enabled some ALS patients to increase their capabilities. Table 1 shows a summary of the characteristics of these three studies. .

Study 1: changes in patients' attitudes toward LTMV[32]

We surveyed 29 ALS patients on LTMV about their attitudes toward LTMV both before and after being put on ventilators. Samples were collected through purposive sampling using personal connections of the researchers. Researchers asked informants three open questions related to LTMV and analyzed their answers qualitatively. We sent questions to 29 patients who were physically capable of communicating and expressing their own feelings and received answers from 20 of them. Though the sample was small and not randomly selected, and therefore difficult to interpret statistically, this survey describes significant aspects of the lives of ALS patients on LTMV that have not been focused on in previous studies.

The results of the survey show that the majority of informants felt that the ventilator had come to feel like a part of their bodies after they began using it, and all the informants who answered remarked that respiratory difficulty was resolved once LTMV was begun (Box 1). Furthermore, many patients reported that their attitudes toward LTMV gradually changed from a vague sense of fear and distrust of the machinery to feelings such as "acceptance", "adaptation", and even "the ventilator is a member of my family".

This change in attitudes occurred mainly through the process of informed consent.

(Kaub-Wittener, et al. (2003)　It assists in patient respiration but cannot prolong life to the extent possible with LTMV

[31] Tracheostomy ventilation. LTMV.

[32] Details of survey 1 are described in Kawaguchi(2006a)

Doctors' positive attitude toward LTMV and families' devoted care and desire for the patients to go on living influenced their willingness to use LTMV to lengthen their lives. Almost all of the informants reported that they did not feel pressured into choosing LTMV by these attitudes of their doctors and families, but rather perceived them positively as welcome support.

Study 2: Achieving well-being in daily life[33]

Is she really happy living like that?

These are the words a passerby said to the attendant doctor of Ms. Hashimoto, a Japanese ALS patient visiting Denmark[34]. Ms. Hashimoto receives LTMV and can only move a few parts of her body. This phrase might represent some people's typical attitude towards living with serious disabilities. Our study, however, showed that some ALS patients on LTMV can in fact enjoy a sense of well-being and do not report a low quality of life. We gathered essays of ALS patients as well as interviewing a few ALS patients to examine how people with this disease can manage to feel a sense of well-being. We picked out four elements as being particularly important to the well-being of ALS patients: being able to relax, enjoying their surroundings (nature, external stimulation), being able to go out, and being able to communicate(Box 2).

Study 3: Active participation in society

In studies 1 and 2, we described the well-being aspect of ALS patients on LTMV. In study 3, we introduce several cases in which ALS patients on LTMV have been able to make significant contributions to society. These patients have realized not only their own daily well-being but have been able to actively participate in society, undertaking a range of activities including studying at a university, organizing a peer-support group, and negotiating with the Japanese government. We conducted a survey of patients who have made full use of Japanese medical and long term care schemes to undertake these sorts of activities. Some of the responses to this survey are listed in box 3 below.

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[33] We based our analysis on a survey done by Tateiwa (2004).
[34] Yamazaki(2006:31)

Discussion

It is clear that the well-being of a patient with advanced ALS, whose physical ability is seriously impaired, cannot be adequately realized without extremely intensive care by others. Many patients and their families are faced with serious difficulty regarding this care and hesitate to employ LTMV. In Japan, however, a number of ALS patients on LTMV have been able to realize a reasonably high level of well-being by utilizing multiple medical and long term care schemes mentioned above and emerging "Independent Living Programs" like Sakura Model.

Certainly, even if sufficient care and treatment are available, it is impossible for ALS patients to achieve exactly the same level of physical freedom or capabilities they enjoyed before their illness. However, as we described, there are examples of ALS patients who positively accept and adapt to their circumstances and enjoy lives different (but not necessarily less happy) than those of people without the disease. We should not simply evaluate such acceptance and adaptation as "poor adaptive preferences". As Nussbaum[2000] argues, this positive kind of "adaptive preferences" is experienced by nearly everyone to some extent .

> We get used to having the bodies we do have, and even if, as children, we wanted to fly like birds, we simply drop that after a while, and are probably the better for it. Again, someone as a child may want to be the best opera singer in the world(as I did), or the best basketball player - but most people adjust their aspirations to what they can actually achieve[35].

This argument can be applied not only to examples of a bird or an opera singer, but also to various problems that are faced by people with disabilities or incurable diseases. Though some reservations are necessary, finding and achieving other functions after losing or giving up some functions should itself be evaluated positively[36]. While to those who have not suffered from this sort of debilitating illness, such as the Danish passerby quoted above, it may seem hard to believe that a reasonable quality of life can be attained when one's physical capabilities have been reduced to the extent they are in advanced ALS patients.

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[35] Nussbaum(2000:137)

[36] For discussions of what kinds of "adaptive preference" should be view positively, see Nussbaum's (2000) criticism of Elster(1983).

Our survey of a group of such patients, however, has found that with a sufficient level of social support and care at least some of those who find themselves in this situation can attain a quality of life that they find high enough to warrant lengthening their lives. .

5. Conclusion

Our surveys indicate that some ALS patients on LTMV have the potential to realize a certain level of well-being when they receive sufficient medical treatment and long term care. This result throws some doubt on treatment schemes that have been constructed, either explicitly or implicitly, on the assumption that a patient of ALS normally doesn't (or shouldn't) want to receive LTMV and LTMV inevitably leads to a low quality of life for both the patient and his or her family.

Leaving aside the controversial issue of what sorts of options regarding the termination of their lives patients should be given, the results of our studies imply that medical and social support systems should not ignore the fact that with sufficient care some advanced ALS patients can enjoy a reasonably high quality of life without placing an undo burden on their families. The question of whether this support can or should be provided is one that then must be faced, and should be considered in discussions of the treatment and quality of life of those suffering from this illness.

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Table 1: Characteristics of the three studies

	Objective	Sample	Method
Study 1	To investigate how patients on LTMV have changed their attitudes toward LTMV.	ALS patients who are sampled by purposive sampling using personal connections of researchers	Email questionnaire
Study 2	To investigate how patients on LTMV are able to enjoy lives, explore their own well-being in daily lives.	ALS patients who wrote essays on the internet or in published magazines or books	Literature review
Study 3	To investigate how patients on LTMV are able to participate in social activities and movements	ALS patients using the SAKURA Model	Case studies

Box1: Selection of comments from Survey 1

Question: What were your feelings towards using a ventilator 1)before receiving a ventilator, 2)just after receiving a ventilator, 3)after some time has passed since you received a ventilator

Selected responses:

- I thought that I didn't want to live if I would need a ventilator. But now I feel that my ventilator is like a part of my family.

- Both ALS and the ventilator are nuisances, but before I knew it, I had accepted or perhaps adapted to my new lifestyle.

- At first, I felt pain because I received a ventilator just after having my ventilator incised but breathing has become really easy and I feel like my body has got lighter. Now the ventilator is a part of my body.

- Machines such as a PC or a ventilator are parts of your own body. To accept this idea is essential for ALS patients like me to broaden our potential and lives.

- Though there are more difficulties than joys, I had never felt as happy to be alive as when I received the ventilator.

- Just after I received a ventilator, I felt as though I had entered a different world. I felt really comfortable and smiled. I found the air incredibly delicious.

- Most importantly I was able to get over the fear of death and became mentally stronger. Receiving a ventilator was less painful than I had imagined. My wife was also glad because caring for me became much easier. I regret not having received a ventilator earlier.

- Even now, I feel that my family needs me and I can advise them about their worries, and I can play an important role in my family. I'm really happy to have received a ventilator.

Box2: Selection of comments from Survey 2

To be able to relax and think

- Anyway, It is not so painful to stay lying down. (text omitted) Rather, I want to enjoy this bedridden time.（Tateiwa[2004:270-271]）

- (Replying to the question whether it is boring to stay lying down at all times) It's not so bad because I have enough time to think（Tateiwa[2004:273-274]）

To be able to enjoy nature and one's surroundings

- The words of the secretary general of Japan ALS Association finally made me decide to receive a ventilator. (text omitted) He suggested that I would able to enjoy seasonal scenery such as cherry blossoms in spring, the sea in summer, red leaves in autumn and snow covered scenery in winter. He also said that there was no telling what sorts of wonderful things I might experience if I stayed alive. He emphasized the joy of living.（Tateiwa[2004:175]）

- A nurse changed the location of my bed. (text omitted) Then a totally different world opened up for me. I saw Mt.Fuji from the window.（Tateiwa[2004:278]）

To be able to Drink

- When tea came into my throat for the first time in the four and half years, I felt the same excitement that I had felt when I used the personal computer that I use now for the first time. I sensed that "my life would be broadened"(Tateiwa[2004])

- A glass of beer was poured into my mouth with a glass needle and entered my stomach leaving a sensation of coolness and fresh bubbles in my throat.（Tateiwa[2004:278]）

To be able to go out

- A major change in my live occurred when I was bought a "portable bird"(portable ventilator). Using this "portable bird", I can go out whenever I want.（Tateiwa[2004:279]）

To be able to communicate

- When half a year had passed since I lost my ability to write, I found that I could communicate by looking at letters on paper and having my wife pick them out. I got to be able to communicate what I was thinking. My life became brighter suddenly (Tateiwa[2004:283])

- I tried to use that(*a communication device) right away. The letters I typed came out as a picture and a voice and could also be printed like on a word processor. I felt that a new world had just opened up for me. This device is really excellent for those who can't physically express their thoughts with their mouth or hands.(Tateiwa[2004:287])

- The feeling of happiness was incomparable when I was able to escape from my passive life by using a word processor that allowed me to type through the movement of my eyelids . (Tateiwa[2004:287])

- The word processor is something like my life itself. It is strange how in a way it is like my heart in that it allows me to continue to be active.

- I will always vividly remember the day I first typed with a word processor the words of gratitude that I hadn't been able to express with a letter board.

Box3: Patients' activities

Peer support

Most patients worry deeply before receiving LTMV, and patients who have already received ventilators consult with them, explain how it is to receive LTMV, and recommend they receive LTMV. Many ALS patients change their minds and decide to stay alive with LTMV after talking to other ALS patients already on ventilators.

Care support business

Some patients on LTMV in the Tokyo metropolitan area have built up their own care business organizations and employ caregivers to whom they themselves train in caregiving. These patients have developed their businesses and sent their own trained caregivers to other ALS patients who begin living home using this home care service.

Social activism

Recently, some patients on LTMV in the Tokyo metropolitan area have gone with their caregivers and other groups representing disabled people to the Tokyo municipal government on an annual basis to negotiate directly about their own care, using letter boards or other communication devices when necessary.

University

An ALS patient on LTMV in Tokyo entered university in 2007 and began studying social welfare.