Decision-Making Process Regarding the Use of Mechanical Ventilation by a Patient with Amyotrophic Lateral Sclerosis: Interaction of the Patient with Family Members and Specialists

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Abstract
Objective: The objective of this study was to examine how specialists should provide support for amyotrophic lateral sclerosis (ALS) patients in the process of their decision-making regarding the use of mechanical ventilation. A case analysis was made focused on the changes in a patient's perception through the process of her interaction with her family members and specialists.
Methods: The author visited the patient's home regularly, and conducted interviews with the patient, her family and the specialists involved.
Results: There were perceptual differences regarding the use of mechanical ventilation among the patient, her family and specialists. These differences arose because of the differences in the patient's perspective, which focused on the present, and that of the specialists, which focused on future developments. Problems resulting from information gaps were resolved by ongoing professional involvement including the provision of information, while problems arising from different values were difficult to deal with.
Conclusion: Specialists should be aware of these differences when offering support. Besides providing accurate information, they should facilitate interaction between the patients and family members, thus helping them to reach a consensus.

Keywords
Amyotrophic Lateral Sclerosis, Interaction, Mechanical Ventilation, Decision-making

I. Introduction

This article discusses the interaction of a patient with amyotrophic lateral sclerosis (ALS) with her family and specialists concerning her decision-making process regarding the use of mechanical ventilation. ALS is a neurodegenerative disease of unknown etiology caused by progressive degeneration of the motor neurons. The disease is usually fatal in two to four years unless mechanical ventilation is used (Society for Prevention and Care Management of Incurable and Chronic Diseases, 2003, pp. 295-6). Two hundred and fifty seven special disease certificates were issued for patients with ALS in Japan in 1974, however, the number increased to 6,646 in 2002 (Society for Prevention and Care Management of Incurable and Chronic Diseases, 2003, p. 446).

Since the practice of informed consent was proposed in the field of medicine, more importance has been placed on supporting patients regarding their choice of treatment methods. The causes of ALS and its treatment methods are still unknown, and quadriplegia, speech disorders, dysphagia, and respiratory problems develop as the disease progresses. However, with regard to speech disorders, communication devices enable patients to communicate, and symptoms management, such as tube feeding for
dysphagia and artificial respiration, help prolong their lives. In 1990 the consultation and management fees for home ventilation were covered by the government health insurance. In 1992 the Medical Care Law was amended, and designated the patient’s home as a place for providing medical treatment. In 1994 when home-visit nursing care programs were expanded to the non-elderly population the care situation concerning patients with ALS also changed. The development of medical science and changes in medical systems increased the choices concerning treatment methods (symptoms management) and the place for treatment, either at home or hospital for patients with ALS, which required them to have the capacity to make decisions.

As ALS patients basically retain their intellectual functions, self decision-making is possible for them. According to Kamo (1) adults who are capable of making judgments have the right (2) to make decisions and to receive information necessary for such decision-making concerning (3) what belongs to them including their body and quality of life, (4) as long as it does not harm others, (5) even if it results in an irrational outcome for the patient (Kamo 1996, pp. 12-3.). Kojima defines self determination as making decisions about their own lives (Kojima 2002, p. 210.). Deciding on mechanical ventilation is literally a choice of life for patients and a difficult one to make. They feel ambivalent between their wish to live and sustaining their lives in a state of immobility.

Many different perspectives have been proposed by professionals to support ALS patients in making the decision regarding mechanical ventilation, but there is little research on whether the patients managed their disease as professionals had expected or how the patients felt about their support. This article discusses how specialists should provide support for ALS patients by clarifying how such a patient changed through her interaction with her family and specialists during the process of deciding on mechanical ventilation, focusing on the perceptual gap of the patient and those concerned with her regarding the use of mechanical ventilation and the changes in the perceptual gap as the key words.

The perceptual gap in the helping process results from the differences in the standpoint of the patient, family members and specialists as well as differences in professionalism among specialists. In fact, professional differences are important in understanding the problems from different angles. However, if the perceptual gap arises when common understanding is necessary in the helping process, problem solving becomes difficult. This research studies the perceptual gap resulting from the different positions of the patient, family members, and specialists in the process of the patient’s decision-making concerning mechanical ventilation and what triggered the changes in their perception.

1. Issues of decision-making capacity and making choices concerning mechanical ventilation

Biestek listed the principle of self-determination as one of the basic social work principles (Biestek 1957-1996, pp. 160-89). According to Biestek, personality develops and matures when people make decisions on their own. However, Kojima refutes that it is taken for granted that self-determination means taking responsibility, but that there are situations in which clients are unable to assume responsibility even if they wish to because of inadequacy or lack of social resources or information (Kojima 2002, p. 230).

ALS patients basically require 24-hour care once they become ventilator dependent. However, in the current situation in Japan, social resources are less than sufficient for ALS patients. There are almost no hospitals or institutions which can accommodate ventilator dependent patients who need long-term hospitalization. Even if patients at home use long-term care insurance, medical care insurance, and all the services available for specified diseases and disabilities they can use only a few hours of care services a day to substitute their primary caregivers, which imposes care burdens on their family almost all day. In addition, in order for ALS patients to choose mechanical ventilation their families are required to have the capacity to take care of the patient at home. In principle, there should be conditions for the patients, which enable them to choose mechanical ventilation or not. However, if their families are incapable of providing care, not a few patients must give up using mechanical ventilation.
Family members must get up two to five times during the night to suck in phlegm in addition to the daytime. They cannot sleep through the night, and accumulate fatigue from care giving. This causes patients to feel ambivalent because they feel that their need for care might change their family members’ lives, while at the same time they have a desire to live. Self-determination under such circumstances is not really self-determination as the patient is dependent on the family’s capacity to provide care.

2. Interaction in the process of decision-making concerning mechanical ventilation

In principle, self decision-making is based on the patient’s autonomy, but various value orientations of the people whom patients interact with and social values influence their decision-making. There are reports that when ALS patients make decisions regarding medical treatment preferences several factors affect the process: provision of information (Young 1994; Moss 1993; Mark 1996), physician’s attitude (Young 1994; Albert 1999; Moss 1993), family (Young 1994; Moss 1993; Mark 1996), attachment to life (Albert 1999), ability to control discontinuation of ventilation (Young 1994; Mark 1996), costs (Moss, 1993), and insurance coverage (Albert 1999; Moss 1993).

Accurate information is essential when patients make decisions about treatment preferences. There are two kinds of information concerning medical treatment and patients’ life with the illness. Physicians play an important role for patients to obtain medical information, and their attitudes influence the patient’s decision-making. According to the treatment guidelines of the Japanese Society of Neurology (Japanese Society of Neurology, 2002) physicians must give patients both positive and negative information concerning mechanical ventilation and give explanations without imposing their own values. "Physicians who had a positive personal attitude toward home ventilation and remained neutral in their presentation of it had a higher percentage of patients on home ventilation" (Moss 1993). Tateiwa states that the basic stance of society should support patients to sustain their lives and that physicians should not take a neutral position but provide patients with a structure which enables them to live, while giving them the freedom to reject it (Tateiwa 2003, pp. 33-6).

Timing is also important concerning the giving of information. "Physicians said that it was best to discuss the option of home ventilation soon after the diagnosis of ALS" (Moss 1993). "Patient preferences may change over time, and clinical education efforts are required throughout the course of disease (Albert 1999)."

Besides information, family and social values are influencing factors. A family’s way of thinking and their relationship with the patient affect the patient both positively and adversely. Many patients with ALS hesitate to depend on mechanical ventilation because of burdening their family members, but in some cases they decide to use it because they feel they are needed by their family. Japanese social values do not approve assisted suicide or stopping mechanical ventilation even if patients wish it. Since they cannot be in control of stopping mechanical ventilation they have no choice to give it a try. This is the main reason why patients hesitate to decide on home ventilation.

As far as social factors are concerned, costs and insurance systems influence the process of the patient’s decision-making. In the United States, “In general, patients in our study had more comprehensive insurance coverage and significantly higher incomes than does average American” (Moss 1993). In Japan, as mechanical ventilation is covered by public health insurance and a policy for developing home medical care services has been promoted, the number of patients on home ventilation has been increasing (Sumida 2002). However, there are gaps between municipalities because of the different attitudes of physicians towards mechanical ventilation as well as the self-help group activities in each area (Shimizu 2001).

The level of a patient’s attachment to life is also an influencing factor. "Patients who stated that they had a strong will to live, had something to look forward to each day, continued to look to the future, etc., were more likely to favor tracheostomy and PEG placement" (Albert 1999).
Decisions on how patients live should be made based on their initiative, but are often influenced by their interaction with their families and specialists. Social values and institutional factors also influence the process. This study focuses mainly on the interaction among the patient, her family and specialists.

II. Methods

Surveys by interviews normally focus on the past feelings of patients. However, the data does not necessarily reflect past feelings but may be affected by present feelings. This research is characterized by its focus on the present feelings of the patient, which arise from physical changes as well as changes in the patient’s environment and discusses the patient’s psychological changes by continuing interviews concerning her present feelings. Secondly, interviews were not carried out with the patient only, but also with her family and specialists. Whenever the physical and environmental conditions of the patient changed, attention was given to her thoughts and feelings, her family and specialists, whether the support was provided as the patient had wished and whether the support provided by specialists was effective were reviewed from the perspective of the perceptual gap between the patient and specialists. Thirdly, in the interim process of analysis the patient and her family were asked to provide input, thus reviewing the support from the perspective of the patient herself. With regard to the choice of a candidate for this research, taking into consideration the characteristics of the research, the patient, who could cooperate with this research from the time of being informed of the disease not long before, and her family were asked to cooperate with this research.

Ms. A (in her 60s) lives with her husband, daughter and the daughters’ child. The family runs a shop from their home, and Ms. A did most of the housework and took care of the shop. There are two more daughters who have their own families. Ms A became aware of her symptoms in 1998, and visited a number of hospitals until she was diagnosed as suffering from ALS at the department of neurology at Hospital A in May 2001. However, she changed hospitals because she lost confidence in her doctor. In July 2002, she was admitted to Hospital B for medical examinations, and her chief physician planned a visit by her to a non-ventilator dependent ALS patient while she was in hospital. In addition, he elicited PEG and tracheostomy since bulbar palsy was progressing more than quadriplegia. The patient rejected both of them, but decided to have PEG at the strong request of her daughter. At the time of her discharge from the hospital a network meeting was held by the staff from a specialty hospital, an Intractable Diseases Information Center, and a health center, and they planned for her to live with ALS at home. At the end of 2002, interventions were begun to elicit her decision-making regarding mechanical ventilation. The following intervention plans were made: home visit by a nurse from the Intractable Diseases Information Center to confirm her and her family’s preferences; visit to a ventilator dependent patient with ALS; and admission to hospital and confirmation of her preferences by her chief physician.

For this research, the author visited the patient’s home regularly to interview the patient, her family and specialists. Since October 2002, the author made visits once or twice a month, asking her and her family if there were any difficulties or changes, and made field notes following each visit. In January 2003, specialists planned interventions regarding her preferences concerning the use of mechanical ventilation. Accordingly, semi-structured interviews were conducted twice for the patient both before and after the intervention and for the family after the intervention. Regular visits to the patient’s home were continued after these semi-structured interviews. In addition, in order to compare the perceptions of the patient and her family by identifying how professionals were involved and from what perspectives, interviews were conducted with her chief physician at the specialty hospital, a nurse at the Intractable Diseases Information Center1, a visiting nurse, and a health nurse (Fig. 1). While the patient was hospitalized the author attended a meeting of the physician and her family with permission from the patient, her family and her chief physician. At the beginning, the author visited the patient with the nurse.
from the Intractable Diseases Information Center, but after the author made an official request to visit the patient for research, they visited the patient separately to distinguish the visits regarding research and care.

Interviews were recorded with consent from the persons who cooperated in the research and a verbatim record was made. The patient answered questions in writing and the author recorded part of the answers while reading them aloud. The author, the nurse from the Intractable Diseases Information Center, and a university professor analyzed the transcript, identified the problems and discussed what kind of support the patient would need. The data concerning the patient, her family and specialists was organized in chronological order and put together to examine the perceptual gap among the patient, her family and specialists and analyzed how their interaction changed the gap. Permission concerning the sharing of data among the team members was also obtained from those who cooperated in the research. The nurse from the Intractable Diseases Information Center continued her actual interventions.

We analyzed the data using the grounded theory approach. Grounded theories create concepts by making comparisons of multiple data. Since this research analyzed the data of several persons who were involved with a single case this is not exactly a grounded theory. However, grounded theory focuses on the process and interaction in the process of building theory. Since this research aimed to discuss the changes in the interaction among the patient, her family and specialists, the analysis using grounded theory seems relevant.

According to Strauss "to bring process into analysis is an essential feature of a grounded theory analysis" (Anselm L. Strauss and Juliet Corbin 1990-1999, p.157); "By process we mean the linking of sequences of action/interaction as they pertain to the management of, control over, or response to, a phenomenon. This linking of sequences is accomplished by noting: (a) the change in conditions influencing action/interaction over time;(b) the action/interactional response to that change; (c) the consequences that result from that action/interactional response; and finally by (d) describing how those consequences become part of the conditions influencing the next action/interactional sequence."

(Anselm L. Strauss and Juliet Corbin 1990-1999, p.143). In the revised M-GAT proposed by Kinoshita he does not fragment data but emphasizes the importance of understanding the context and examines the person's perception, action and emotions reflected in the data as well as related factors and conditions (Kinoshita 2003, p.158).

This research examines the process of changes in the patient's psychology following the course of time from the onset of her disease, and its diagnosis to her decision-making regarding preferences concerning mechanical ventilation and it is considered that evaluating the effectiveness of professional support from the viewpoint of the patient is possible by investigating whether the outcomes (changes in the patient and her family) have been obtained as specialists planned, as a result of the changes in the conditions produced by the support of specialists.

In publicizing the data analysis, the contents were confirmed and approved by the patient and her family.

III. Results

Four categories were identified as the cause of the perceptual gap in the process of decision-making by the patient regarding the use of mechanical ventilation. As far as her relationships with specialists are concerned, there was a gap between the information which the patient and her family desired and the information provided by specialists. As for the family, there was no sharing of the issues by the family members. There was also a perceptual gap between the patient and her daughters arising from their mother's perception of her disability, which she did not want to expose to other people. In addition, there was a perceptual gap among the three parties; the patient, daughters and specialists, in terms of their perspectives concerning the present and future. The following is a detailed discussion of the interaction among the patient, her family and specialists in these four categories.
1. Information desired by the patient/her family and information provided by specialists

Three years elapsed until Ms. A was diagnosed as suffering from ALS since she began to feel that something was wrong with her tongue. The perceptual gap during this period between the patient and her family was that, while the patient was seriously worried about the symptom, her family did not take it seriously. In January 2001 she saw a neurologist at Hospital A and was hospitalized in May for medical examinations. Her family, who had not taken it very seriously, was totally perplexed when the patient was unexpectedly diagnosed as suffering from an incurable disease. They were even more confused because of the way the physician informed the patient of the disease. In spite of the serious matter of life or death for the patient, her physician gave her a brief one-sided businesslike notice, while ignoring her family’s feelings. Furthermore, the provided information was insufficient, with no specific detailed explanation about mechanical ventilation. He only mentioned that he did not recommend it because it involved major burdens.

The patient’s daughters gathered information from the Internet because they had no knowledge about the disease or the life of ventilator-supported patients. They learned from the Internet about the approach to inform patients of the disease step-by-step and asked the physician to follow the step for their mother. However, the physician only offered a choice of either informing her of the diagnosis or not. He did not give a detailed explanation to Ms. A. After all, her daughters explained to their mother about the disease and mechanical ventilator support. Almost no information was provided by the physician either when Ms. A went to see him for an examination as an outpatient. Because of the gap between the information provided by the physician and the information which the patient and her family wanted, they lost trust in the physician and began to look for a good hospital. They wanted to find a hospital which would provide good emotional care in helping Ms. A decide whether to use a mechanical ventilator or not.

Following their search the family moved Ms. A to Hospital B, which had a good reputation. This hospital provided medical information according to the patient’s condition as well as information on living with ALS. When Ms. A was hospitalized for examinations at Hospital B her new physician found the need of ventilatory support for her in the near future because of the progression of bulbar palsy. He first confirmed the family’s preferences concerning the use of mechanical ventilation and gave a detailed explanation to them and the patient. The physician mentioned that he had explained briefly to Ms. A about mechanical ventilation and that her initial response was “I do not want to live (with ventilatory support)” and “I can finish now.”

Ms. A’s chief physician at Hospital B arranged a visit with an ALS patient for Ms. A, her husband and daughters to provide them with information on living with ALS. The ALS patient had bronchotomy but refused mechanical ventilatory support. The patient’s four limbs were mostly paralyzed and needed total care. Her husband, who was taking care of his wife, said in her presence “I cannot sleep at all.” This gave Ms. A’s husband an impression that the care would be a heavy burden. The nurse from the Intractable Diseases Information Center said that Ms. A’s husband probably had an image of home ventilation as equal to attending to all her personal needs and that he would not be able to do it. One of the daughters living separately from their parents took positively what the husband of the patient they visited said. She found it necessary to have a relationship in which caregivers can feel free to say anything and accordingly that the patient does not feel hurt by that. Similarly, regarding the information on the life of the patient with ALS they visited, Ms. A’s husband had an impression of providing care as a heavy burden, while his daughters had a strong impression of the family whose relationships were so open that family members could say anything to each other. Thus, there was a gap in the way they processed the same information.

When Ms. A was discharged from Hospital B a framework which would make ongoing care possible both at the specialty hospital and her home was structured. The information obtained by the nurse from the Intractable Diseases Center, the visiting nurse and the health nurse during their visit to the patient at home
was delivered to the patient’s chief physician at the specialty hospital, and the hospital in her neighborhood and the specialty hospital were collaborating with each other to deal with emergencies. The nurse from the Intractable Diseases Center was working closely with the patient’s chief physician at the specialty hospital and its ward nurse and shared information also with home-care service providers (Fig. 1).

2. Sharing of the issue by the family: interventions in the process of self decision-making for home ventilation

Ms. A, who was running a shop with her husband, felt uneasy with her family when she was not able to work any longer. She began to sleep in a separate room from her husband because she did not want to disturb him when he was tired from the day’s work. During that period her bulbar palsy progressed, making verbal communication almost impossible, and there was a danger of choking with phlegm stuck in the throat. Because her consideration for the family might lead to an emergency the visiting nurse urged the couple to sleep within an area in which she could reach out her arm to give her husband a signal if an emergency arose like choking with phlegm. The nurse also told Ms. A that in the event that a worst situation occurred because she was not able to wake him up, he would be the one who suffered remorse.

Later when Ms. A’s swallowing function began to decline, an accident happened. Her tablets got stuck in her throat. The visiting nurse asked Ms. A’s chief physician at the specialty hospital to confirm her preferences concerning the use of home ventilation to prepare for emergencies. Since Ms. A’s hospitalization had been planned to change the gastric cannula her chief physi-
cian at Hospital B decided to take the time to discuss the matter with Ms. A while she was hospitalized. In order to provide her with information regarding the ventilator dependent life he arranged a visit to a ventilatory supported patient with ALS before Ms. A was hospitalized.

During her first hospitalization at Hospital B when she was given an explanation about mechanical ventilation, she revealed her preference not to use it for the first time. Her daughters told their mother that they wanted her to use it, but she refused. Since then Ms. A and her family stopped talking about it. For some time, after she was discharged from the hospital, the topic was taboo for the family. However, due to the interventions made by the specialists, the patient and her family began to think individually about mechanical ventilatory support, but did not share the issue among themselves. Ms. A did not confide her worry to them either.

The nurse from the Intractable Diseases Information Center, inspired by the visiting nurse and the interim analysis of this research, began to make interventions to help the patient make her own decision. To begin with, she confirmed with Ms. A and her family at their home concerning her preferences with respect to mechanical ventilation. Her daughters wanted their mother to use it and were prepared to support her once she decided to do so.

When the specialists began to be involved in her decision-making process Ms. A's husband showed more interest in the non-use of a ventilator. He asked the following questions to the author in front of his wife: “I want to hear what the caregiver of a patient with no ventilatory support has to say” and “Would she suffer more when she is dying if she was not supported by the ventilator?” When her husband was not with her Ms. A said, “My husband does not want me to use a ventilator.” However, her husband said when he was not with her “Maybe, it would be best for her to use it.”

Her husband, who had a strong impression of heavy care burdens when he and his family visited a patient with ALS for the first time, was not positive about home ventilation because he thought that, based on her personality, his wife would not need it. He also said at the beginning that he did not know much about ventilators. However, basically, he did not show much interest in getting information since he did not have much time to read all the materials his daughters gave him. His knowledge about the disease and care was biased. There was a gap in his perception of the disease with that of his family, and he had little interest in it.

The second patient with ALS, who Ms. A and her family visited, originally did not want to use mechanical ventilation, but was persuaded to do so by her family. When Ms. A and her family visited the first patient before this patient, Ms. A's husband had a strong image of heavy care burdens. Therefore, an ALS patient who still had some mobility was chosen for their second visit. The caregiver of this patient said to Ms. A and her family that it was good that she decided to use a ventilator, and the patient gave a smile. Ms. A felt that the caregiver’s attitude must be a big relief for the patient. As Ms. A's husband’s image of ventilatory-life changed through his observation that the ventilatory-supported patient seemed to be moving forward with enthusiasm and that her facial expression looked brighter than the patient without such support, he began to think that he would regret it if he did not let his wife use it.

After the visit to the patient with ALS, Ms. A's husband asked his wife if she wanted to use mechanical ventilation, but it was a one-sided question from him. As a result, there was no clear response from her. However, when her husband saw a mechanical ventilator for the first time he was encouraged by the nurse from the Intractable Diseases Information Center to ask a question, and his question changed to a more specific one about her actual ventilator dependent life. The nurse from the Center asked one of her daughters if she had ever imagined her mother’s ventilator dependent life, after which the daughter actually began to have a clearer image of the placement of her mother’s bed and furniture. Ms. A said she had not discussed home ventilation with her following their visit to the ventilatory-supported patient but said that she had heard from her husband that their daughters were talking about buying a car which could accommodate her wheel chair.

When the specialists began to be involved with the process of Ms. A's decision-making one of her
daughters, who was living away from home, encouraged her mother to express her preferences regarding home ventilation. When the family visited the first patient with ALS, the daughter felt that her family should share the issue among themselves and thought that the topic should not be special so that they could talk freely about it. When she visited her parents she asked Ms. A about home ventilation while they were carrying on an ordinary conversation. At that time the question was posed only from her side, and there was no response from Ms. A. However, this led her to express her preferences to her daughter who lived with her. “When she (daughter living separately) asked me about the ventilator I was not able to answer properly, but I want to live.” Actually, Ms. A decided to use mechanical ventilation when the specialists began to make interventions and her physical conditions were not as good as they were before the family visited the second ALS patient. After her visit to the second patient she said, “That was what I had imagined. Nothing changed (in terms of her feelings).”

During her second hospitalization her chief physician at Hospital B met with Ms. A to confirm her preferences concerning mechanical ventilation. During the meeting Ms. A said, “I want to use the ventilator as long as I can remain mobile.” Her feelings changed from the first time she expressed her preferences. Since Ms. A’s bulbar paralysis was progressing more rapidly than that of her four limbs it was quite likely that she would use the ventilator while she remained mobile. However, the question remained as to what should be done later when her four limbs became immobile. Ms. A said the possibility of using the ventilator would be fifty percent if she became immobile.

At the meeting with the specialists and the family, they tried to share Ms. A’s preferences. However, during the interview at a later date it was found that Ms. A’s daughters and her husband did not confirm her preferences directly with her. Also, after the meeting, the nurse from the Intractable Diseases Center received the information from a ward nurse that Ms. A was doubtful regarding what her family had said. In order to coordinate with her family, the nurse from the Center obtained permission from Ms. A and confirmed with her family concerning their preferences at home. Her husband said “I would regret it if my wife died without mechanical ventilation, so I want her to use it.” Her daughter who lived with them said, “I want her to continue to live” because she felt lonely when her mother was in hospital. The nurse from the Center told Ms. A that she was needed by her family. Ms. A held the nurse’s hands and cried.

3. Perception of disability

Ms. A’s perception of her disability, which she did not want exposed to other people, had a major impact on her understanding of the disease, and her husband had a similar perception. Since Ms. A found it difficult to communicate with other people she hated to expose herself to them and did not want to be talked about. As a result, she began to withdraw herself and said, “If I must depend on a mechanical ventilator I want to go to a place that nobody will know” and also said “I want to die (in obscurity) like an elephant.” Her daughters felt ambivalent because they wished their mother to live longer and at the same time were uncertain whether she would be truly happy to depend on ventilatory support, given her perception of disability as a disgrace as well as her feelings about continuing to live while remaining immobile. They left the decision to their mother and did not push her to make her own decision concerning medical procedures, considering that she would not be able to manage the situation unless she had a will to live and that they would accept it if she decided against the use of mechanical ventilation.

There was a gap in the perception of disability between Ms. A and her daughters. Ms. A did not want to expose herself in a wheelchair to other people, while her daughters wanted her to go out even with ventilatory support. Her daughters also wanted her to continue to have contact with the community and wanted her neighbors to help her when she was on mechanical ventilation. They thought unless she could live peacefully surrounded by her neighbors her ventilator dependent life would be depressing. They were anxious about their mother who could not be open about herself. When the whole family discussed mechanical ventilatory support her daughters told their mother that they were worried
about her using the ventilator, while being unwilling to expose herself to other people. The daughter who lived separately said to her mother as she was leaving for another room while her husband was visiting the house, “There’s no meaning in using the ventilator if you do not find the joy of watching your grandchild running around the room while you are lying in bed. That will make you feel worse.” However, Ms. A did not respond to her daughter, and it ending up as a one-way communication. The nurse from the Intractable Diseases Center felt the distance between Ms. A and her family.

When Ms. A and her family visited the second patient with the same disease her husband said honestly to the caregiver of the patient that he did not want to expose his wife’s disability to other people. The patient’s caregiver said that nothing would begin unless they accepted the reality. The nurse from the Intractable Diseases Center felt that the caregiver made a difference for Ms. A and her husband, who did not want the disability to be exposed. The caregiver’s comment triggered change in Ms. A’s husband’s perception. Later he said: “I would be open about the situation if my wife became bedridden” and “I won’t be able to manage this unless I let it all out into the open.”

According to the visiting nurse, the facial expression of the daughter living with her parents looked gloomy when the nurse began visiting the family and she would reconfirm several things with the nurse concerning the medicine prescribed to her mother, but after the meeting her face took on a bright look and she no longer reconfirmed about the medicine. According to the health nurse, Ms. A used to express herself as a source of trouble for her family and did not express her wish to live, but after the meeting she began to say she wanted to live. Early on, Ms. A’s husband did not appear himself even if he passed the room where the health nurse was visiting his wife, but after Ms. A’s second hospitalization, he changed and spoke to the health nurse asking, “How are things?”

Her husband used to say, “I don’t think my wife will use ventilation,” but his attitude was different after the meeting when his wife was hospitalized for the second time. He said: “I would regret it if she did not use a ventilator,” and “Nobody wants to die.” However, his daughters felt that their father had said he wanted his wife to use mechanical ventilation because he was overwhelmed by his emotions, while they were still ambivalent about the use of ventilation because of their mother’s perception of disability.

4. A perspective focusing on the present and a perspective focusing on the future

When Ms. A entered Hospital B for examinations the first time, she was recommended by her chief physician to have tracheostomy and PEG, but she refused to have both of them. Her family was still unable to make their final decision concerning mechanical ventilation, however, as they saw her becoming physically weak and losing weight, the daughters suggested PEG, expecting her to maintain her energy by feeding nutrition into the stomach. Thus, they encouraged her to make a decision regarding the medical treatment procedures. However, they did not push her to make a decision regarding the use of mechanical ventilation as she refused to use it. Right after Ms. A began to receive home care she and her family did not touch upon mechanical ventilation. Since she was able to speak and to write she was given an explanation about an apparatus for communication, but she did not practice using a personal computer.

The visiting nurse assumed that there was another barrier to tackle concerning the use of home ventilation. She thought it would be impossible for Ms. A to accept her disability before she actually faced the reality of her situation. She considered that Ms. A would overcome the barrier by facing the reality, and also that it was important for the family to wish for her to survive by all means and that their feelings must synchronize with her wish to live with her family.

IV. Discussion

1. Issues concerning interaction among family members

Ms. A’s daughters expressed their willingness to support their mother in the event she chose home ventilation. However, she had not been able to express
her wish to live.

It is said that families and social values affect the patients’ decision-making regarding the use of mechanical ventilation. The primary issue for the family in this research was that the family members did not share the problems resulting from the patient’s illness with each other. Despite the family’s critical situation Ms. A had not expressed what she had in mind to her family. Her daughter who lived separately from her parents felt that unless the family members could say openly to each other what they thought, they would not be able to face this kind of difficulty and she wanted her parents to be open with each other. She wanted to build a family relationship, which enabled them to talk about this kind of problem in their daily conversations.

The second issue arose from the couple’s perception of disability. Both husband and wife had a strong feeling that they did not want their neighbors to know about her illness. Ms. A said she wanted to die (in obscurity) like an elephant. They were still influenced by the old social values that prevailed in the days when persons with disabilities were not able to go out of the house of their own free will.

The daughters left the decision to their mother because they were uncertain whether she would be happy to use mechanical ventilation considering her perception of disability and because she did not want to be exposed to other people’s eyes. Ms. A at first refused to have PEG but finally agreed to have it with her daughters’ encouragement. However, there was no push by her daughters about her preferences concerning mechanical ventilation. It is often difficult to make a decision without a push by the family, as the patient must depend on his/her family’s care to live with mechanical ventilatory support. Social values which impose responsibility on the patient for their self decision-making affect the family. In this regard, Ms. A’s daughters might feel that they could not take full responsibility for the possibility that their mother might regret her choice regarding home ventilation. Furthermore, her husband did not push her to choose it either. However, not a few patients have chosen home ventilation because their spouses took the illness seriously and assumed the initiative, saying, “This is tough but let’s work together,” and, “I want you to live.” Ms. A’s husband thought initially that his wife did not want to use a ventilator, and his wife, sensing his feeling, thought that he did not want her to use it.

One of the key elements in facilitating self decision-making is said to be the family. Ms. A’s family strongly wished her to live. However, since her perception of disability and not wanting to be exposed to other people’s eyes did not change, her daughters’ ambivalent feelings remained unchanged.

2. Interventions by specialists concerning information

It is said that physicians should give patients medical information in a neutral and fair manner including the merits and demerits of using mechanical ventilation in stages as early as possible. It is also said that they need to explain to the patients that the care will be provided on a continuing basis, and that depending on their preferences an explanation regarding palliative care will be necessary. Ongoing education is necessary because patients waver emotionally.

The physician at Hospital A expressed his opinion that he did not recommend mechanical ventilation before giving a detailed explanation of it to the patient. There was a big difference between his explanation and the information the patient and her family wanted. The physician’s attitude aroused the family’s mistrust in him. After the patient and her family received an explanation at Hospital A, her family asked for more information about the illness and other information which would help them to have a clear picture of her life after she was put on a mechanical ventilator and also asked for ongoing care. The physician’s attitude in giving medical information and the amount of information provided were very different between hospitals A and B. Hospital B arranged a visit with another patient with the same disease for the purpose of providing information about the patient’s life at home, and when the patient was going to be discharged they organized a social network or ongoing home care with the specialty hospital and the Intractable Diseases Information Center. It is said that an explanation concerning palliative care is also necessary in this situation, but in these
hospitals there was little information on palliative care for the patients who do not choose mechanical ventilation. Ms. A's husband asked the author about death in the event his wife did not choose mechanical ventilation. Also, he wanted to hear from the caregiver of the non-ventilatory dependent patient about his feelings. It seems necessary to give information concerning the merits and demerits of using mechanical ventilation as well as its non-use from the perspective of the patient's family. Particularly, in the event that the patient does not use it, it is quite likely that their family will regret it. It is obvious that the patient will need palliative care if they refuse mechanical ventilatory support, while at the same time grief therapy will be necessary for the family.

Because each individual has his/her own sense of values it is quite natural that people have different perceptions of the same information. After the first visit to the patient with the same disease, the daughter living separately from the family appreciated the relationship between the caregiver and the patient in which they could say openly to each other what was on their mind, positively. Ms. A's husband was struck by the heavy care burden and said he did not think his wife would use a ventilator. Ms. A sensed her husband's feelings, which probably prevented her from expressing her wish to live. There was a small gap between her husband's perception of the disease and that of the specialists and his family. However, his perception of the disease changed through the interventions by the visiting nurse and the nurse from the Intractable Diseases Information Center, but his and his wife's perception of disability did not change. Therefore, their daughters wondered if their mother would be really happy to have mechanical ventilation, feeling that she did not want to expose herself to other people's eyes, which was rooted in her perception of disabilities. As a result, the ambivalent feelings of her daughters remained unresolved. It is possible to bridge the gap in the perceptions of the information given to the patient and her family through continuing education by specialists and to solve the problems which resulted from the gap. However, it is not easy to change their values.

It is necessary to give information while taking into consideration the relationships among medical care practitioners, patient and family. At the beginning Ms. A and her family avoided the topic concerning mechanical ventilation and each of them was worried about it individually. The nurse from the Intractable Diseases Information Center served as a coordinator by listening to what Ms. A and her family had to say together as well as listening to them individually. In addition, arranging visits for Ms. A and her family to patients with the same disease and providing them with opportunities to talk with the physician about mechanical ventilation during her hospitalization, led her and her family to seriously consider its use. Dependence on mechanical ventilatory support means imposing care burdens on the patient's family. It is difficult for the patient to make decisions if the family members cannot share the issues with each other and reach a consensus even if information has been given to them individually. Specialists need to facilitate interaction among family members besides providing them with information.

One of the factors which make the patients' decision-making regarding mechanical ventilation difficult is that they must decide not about the present situation but about their future crisis. Particularly, at the stage at which their disability has not yet turned worse they find it difficult to accept the information that they will need ventilatory support in the future. They may be able to absorb it only when they face the crisis. Specialists urge the patients to make decisions by providing them with information in advance in order to formulate appropriate measures for the future crisis which is inevitable. There is a major gap between the perspective of patients for whom their future is inconceivable, as they are so overwhelmed by their present condition that they do not want to see what the future holds, and that of the specialists who want to offer support by focusing on the future. For patients with ALS, it is said that continuing education appropriate for each progress of the disease is necessary. However, unless support is offered, while recognizing the gap between these perspectives, the gap of perceptions between the patients and medical care practitioners will remain unchanged.

This article has discussed professional support focusing on the gap of perceptions as its key words.
Since this research deals with only one case, the author must be prudent in concluding its outcome as the concept of support for patients with ALS in general. In addition, this article deals with a patient who had little contact with the ALS patients support group and other patients with the same disease but was mostly involved with specialists. Therefore, the author plans to make comparative case studies of patients who have more contact with other patients with the same disease and patients who have little contact with specialists.

V. Conclusion

One of the factors which make the patients' self decision-making toward mechanical ventilation difficult is the social factor that the decision depends on the capacity of their families to provide care and that it is a decision not about the present situation but about the future crisis. There is a big gap between the perspectives of the patients who have not accepted their present reality or do not want to see what the future holds and those of specialists who try to support them with a focus on the future. In this regard, support must be provided while recognizing these gaps.

Educational information and the attitudes of physicians play important roles in the decision-making process of patients. Information should be provided focusing on mechanical ventilation as well as the merits and demerits of its use and non-use. However, it should be given both from the perspective of the patient and their family. Sometimes family members do not share the problems with each other even if educational information has been provided to them and the patient. Specialists should give support to facilitate interaction between the patient and his/her family members and encourage them to reach a consensus, not simply provide information.

In the case of Ms. A some of the problems, which resulted from the gap of perceptions concerning information, were solved through the continuing involvement of specialists. In order to be able to accept disability, patients need to change their values (Wright, 1960), but in this particular case it has not happened yet. Because of this the problems arising from the different values remained unsolved. Exploring an appropriate form of support was the immediate challenge so that the patient would not regret her decision to use mechanical ventilation.

Notes:
1) The Intractable Diseases Information Center is engaged in the promotion of comprehensive home medical care practices including medical care practices at home, consultation, training for medical care practitioners and research, with the main focus on improvement of the patients' care environment through enhancing the coordination of institutional care and home medical care for intractable disease patients.

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