J Neurol (1998) 245 : 717–722
© Springer-Verlag 1998

ORIGINAL COMMUNICATION

Gian Domenico Borasio
Raymond Voltz

Discontinuation of mechanical ventilation in patients with amyotrophic lateral sclerosis

Received: 24 November 1997
Received in revised form: 3 April 1998
Accepted: 5 April 1998

G. D. Borasio (✉) · R. Voltz
Department of Neurology,
University of Munich,
Klinikum Grosshadern,
Marchioninistrasse 15,
D-81377 München, Germany
e-mail: Borasio@lrz.uni-muenchen.de
Tel.: +49-89-7095-3671
Fax: +49-89-7095-3677

Abstract Mechanical ventilation, both invasive and non-invasive, may be an effective means of improving the quality of life and prolonging the survival of patients suffering from amyotrophic lateral sclerosis (ALS). However, the attitude towards this palliative measure varies greatly between different centres and countries. One of the arguments cited against this procedure is the fear that a patient might request the physician to discontinue life support. We believe that the question of withdrawal of mechanical ventilation can only be meaningfully addressed in the general context of palliative care. Here, we review possible modes of action in response to a patient's request for life support withdrawal and their medical, legal and ethical implications. We propose that the following goals should be pursued: (1) prevention of unwanted ventilation by early, open discussion with patient and relatives, (2) delivery of optimal palliative care by the caring team, (3) recognition of the patient's right to withdraw his/her consent to an invasive medical procedure. If these goals have been met, it may be medically, legally and ethically justified for the physician to take all necessary steps to ensure a peaceful death after discontinuation of life support.

Key words End-of-life decisions · Life support withdrawal · Tracheostomy · Palliative care · Motor neuron disease

Introduction

Amyotrophic lateral sclerosis (ALS) is a degenerative disorder of the motor neurons that leads to a progressive weakness of voluntary muscles, while clinically sparing all other parts of the nervous system [9]. Death usually ensues within 3–5 years of onset, from insufficiency of the respiratory muscles. Mechanical ventilation has been increasingly recognized as an effective means of improving quality of life and prolonging survival in patients with advanced-stage ALS [19, 25]. Two major types of ventilatory support must be distinguished. Non-invasive, intermittent ventilation via mask serves the primary purpose of palliating the symptoms of chronic hypoventilation and is usually administered at night. Invasive ventilation via tracheostomy, on the other hand, has a life-prolonging effect, and the patients usually require uninterrupted ventilatory support. The employment of mechanical ventilation, both invasive and non-invasive, varies greatly between centres and countries, depending on the physicians' attitudes [20, 21]. One of the controversial aspects of this procedure is the issue of life support withdrawal at the patient's request [15].

In the following, we report two cases of invasive ventilation in which such a request was made, then discuss the physician's possible reactions to this request and their medical, legal and ethical aspects. We will take the situation in Germany as an example of the legal considerations involved. It differs, for example, from the liberal legislation in the Netherlands, but is similar to that in many other European countries and in parts of the United States, where legislation concerning terminally ill patients is absent [15]. It is not the intention of this article to provide a

comprehensive overview of existing legal practices, but rather to stimulate further discussion on this topic.

Case reports

Case 1

This 57-year-old ALS patient had repeatedly asked not to be intubated in the case of terminal respiratory insufficiency. However, the patient's own son, himself a physician, initiated artificial ventilation when his mother became unconscious owing to respiratory failure. The patient was hospitalized in an intensive care unit. Using a typewriter, she asked the hospital doctors to end ventilation. When they refused, she asked her husband to disconnect the respirator from the power supply, which he did during the night. Before the hospital staff realized what had happened, the patient died. Her husband was put to trial under charge of "killing upon request", a crime according to German law. His eventual acquittal was the source of much debate; the case was discussed on nationwide television in the popular legal series entitled "How would you decide?". Most of the spectators, through a telephone poll, thought that the acquittal was correct, but a strong minority (around 25%) thought that he should have been found guilty and punished.

Case 2

This 67-year-old patient was diagnosed with ALS as an inpatient in a neurology department. He was told that he had an incurable and progressive neuromuscular disease. He was given an appointment in the outpatient clinic to receive further information, but did not come. A year later, respiratory failure developed. The patient's wife called the emergency physician, who intubated the patient and transferred him to the neurological intensive care unit, where tracheostomy was performed. Three weeks later, home ventilation was initiated and well tolerated by patient and family. Five months later, the wife called the hospital and reported that her husband had repeatedly expressed the wish to have the ventilation discontinued. In the doctor's presence, the patient repeated his wish and asked the doctor to take all necessary steps to ensure a peaceful death. At this point, there were no doubts about the diagnosis; the patient was in full possession of his mental faculties, but physically unable to stop the ventilation by himself.

Comment

In case 1, the patient was informed in advance of the impending respiratory failure and had expressly refused terminal intubation. This situation is still not always the case, since many physicians in Europe do not consider it appropriate to discuss all aspects of a disease such as ALS with patient and relatives. Frequently, the patient is given reassuring statements, while the relatives are told separately that the patient will die of the disease within 2–3 years and that "there is nothing that can be done to help him". The logical consequence of this attitude is that many patients are not informed about the final stage of the disease and never have the chance to express their will regarding life support measures. However, withholding information and treatment options can give rise to exactly the situation that the physician is trying to avoid, as exemplified by the second case [7]. Similar cases have been reported in the literature [13, 15, 32] and anecdotally among colleagues numerous times, mostly involving patients who had been intubated in an emergency situation without knowledge of the diagnosis. In the following, we would like to discuss – without any claim to completeness or objectiveness – some of the physician's possible reactions to a patient's request to remove the ventilator, together with the medical, legal and ethical implications. The possible medical responses to the patient's request for ventilator withdrawal can be summarized as follows:

1. Complete refusal and continuation of life support
2. Discontinuation denied, but no therapy for complications (e.g. no antibiotics in the case of pneumonia)
3. Compliance with the patient's will: three possible modes of action
   a. Discontinuation by the patient without any action by the physician
   b. Discontinuation by the patient with subsequent sedation
   c. Sedation with subsequent discontinuation by the physician

Medical and legal considerations

1. Complete refusal and continuation of life support

Medical aspects: This is not an uncommon response. Many physicians believe that they have a deontological (i.e. a priori) professional duty to preserve life, regardless of the circumstances. In addition, fear of potential legal consequences can also lead to a complete disregard of the patient's wish (see case 1).

Legal aspects: (The legal considerations outlined here are based on the published expertise of Prof. Dr. jur. H. Schöch, University of Munich [22]. In view of the absence of relevant legislation and the few court rulings in Germany concerning this subject, future court decisions are not bound by the principles outlined.) According to the principle of patient autonomy, every invasive medical procedure requires the patient's informed consent. If con-

sent is withdrawn, ventilation becomes an unwanted, and thus unwarranted, treatment. Thus, ventilating a patient against his/her explicit will could lead to a damage suit or even a penal trial of the physician (on charges of battery/trespass against the person) [5].

2. Discontinuation denied, but no therapy for complications (e.g. no antibiotics in the case of pneumonia)

Medical aspects: This "pragmatic stance" is a frequently taken, tacit compromise: the physician refuses to take any "active role" in the patient's death, but does not want to "unnecessarily prolong life". However, it must be noted that, especially in young ventilated ALS patients, life-threatening complications such as pneumonia may take months and even years to occur [16].

Legal aspects: As regards the doctor's refusal to end ventilation, the situation is similar to the previous one. The non-treatment of complications entails no legal consequences if it is at the patient's request.

3. Compliance with the patient's will: three possible modes of action

a. *Discontinuation by the patient without any action by the physician*

Medical aspects: In this (theoretical) possibility, the patient would shut off the respirator himself, if necessary using a specially constructed myoelectrical switch or other electronic device. The physician would not take any action either before or after disconnection. This would cause the patient to go through a phase of severe, unrelieved terminal dyspnoea before dying.

Legal aspects: This mode of action could be punishable by law, because the physician fails to take any appropriate action (e.g. morphine or sedation) to relieve the patient's terminal dyspnoea after disconnection.

b. *Discontinuation by the patient with subsequent sedation*

Medical aspects: Sedation can be used as a palliative measure to prevent terminal dyspnoea. However, if sedation is initiated after life support withdrawal, the timing can be crucial. Two dangers must be avoided: too light a sedation might not relieve dyspnoea, while too high a dose might actually kill the patient before the hypoxia does.

Legal aspects: This procedure is not punishable per se, but may entail some risks. If sedation after life support withdrawal is too light, the physician might be guilty of insufficient palliative treatment. On the other hand, if the sedation is too strong and kills the patient before hypoxia ensues, this may be regarded legally as manslaughter in some countries. We believe that the principle of "double effect" [2, 17] would justify the use of as high a dose of sedative as is deemed necessary by the physician, as long as it is not administered with the intention of killing the patient. However, this principle is not yet universally recognized as legally binding.

c. *Sedation with subsequent discontinuation by the physician*

Medical aspects: In this case, sedation is employed as a preventive palliative therapy for terminal dyspnoea. Sedatives can be administered safely while the patient is still being ventilated. This approach has been advocated by several authors [10, 14]. However, it requires that the physician himself takes all essential steps, including the disconnection of the ventilator. A possible sedation regimen employing benzodiazepines and morphine is shown in Table 1. Barbiturate administration before disconnection has also been described [29].

Legal aspects: This procedure is possibly the safest method from the juridical point of view. It may be argued that the patient, once sedated, is not "in control" of the situation any more and that any theoretical change in his/her will would not be detected. However, the patient clearly has the power to consent to the disconnection procedure *as a whole*. Thus, the consent of the patient to the sedation as part of the life support withdrawal procedure does not end with the onset of unconsciousness (in this respect, the situation is legally not dissimilar to a routine surgical operation under general anaesthesia). In addition, since sedation in this situation is administered to prevent dyspnoea, and not to kill the patient, it would also fall under the definition of "double effect" discussed above.

Table 1 Possible regimen for conscious patients requesting sedation for ventilator withdrawal (from [10], with permission)[a]

Indication	Treatment
Before withdrawal	Bolus dose of 2–4 mg of midazolam
Distress during weaning	Bolus dose of 5–10 mg of morphine, followed by continuous morphine infusion (50% of bolus dose/h)
Further distress	Repeat bolus dose; increase infusion rate correspondingly

[a] These doses are for patients who were not previously taking anxiolytic drugs or opioids; if a tolerance for these drugs is established, higher doses will be needed

Ethical aspects

From what has been said, the decision-making process might seem relatively straightforward, if one considers only the medical and legal lines of reasoning. However, none of the above is set in stone. We are aware of the limitations of any theoretical approach in matters of life and death, and we believe that the ultimate decision should also be based on ethical considerations, which may overlap or conflict with the medico-legal arguments, depending on the individual case. As an example of a possible way to put ethical principles into practice, the "proposal for a new legislation concerning assisted death" [3] was formulated in 1986 by a group of German medical and law scholars (Table 2). A comprehensive discussion of all ethical questions posed by such extreme cases is beyond the scope of this article (for review, see [12, 26, 31]). Nevertheless, a few considerations may be permitted.

In Europe, traditional Christian deontological ethics stresses the intrinsic value and dignity of each single human life per se, i.e. regardless of the actual situation. This ethical view considers life support withdrawal as equal to active euthanasia. On the other hand, teleological (i.e. context-oriented) ethicists agree that there is no general obligation to prolong life regardless of the circumstances. However, there is also a general agreement that the patient's will cannot be the sole valid basis for the ethical decision [11]. The physician has to balance the patient's will against all aspects (positive and negative) of the patient's conditions of life and the presumable future course of the disease. The ethical principle of patient autonomy must be weighed against the principles of beneficence (acting in the patient's best interest), nonmaleficence ("above all do no harm"), fidelity (faithfulness) and justice [4, 12]. Owing to the complexity of such a decision, the advice of all members of the caring team (relatives, physician, nurse, psychologist, hospice team, religious ministers, etc.) is required. However, the final responsibility still resides with the physician.

Table 2 Proposal for a new legislation concerning assisted death [3] (excerpt)

Principles of the proposal

- Protection of life, not an obligation to live: protection of the patient's wellbeing, his right to self-determination and his human dignity in the last stages of life
- All decisions must be based on the patient's will and situation
- "Help for the dying" is preeminent over "help to die"
- *in dubio pro vita.*

§ 214 Discontinuation or non-initiation of life-supporting measures

He who discontinues or does not initiate life-supporting measures does not act unlawfully, if

1. The person concerned has explicitly and earnestly requested this action.
2. …

Discussion

Mechanical ventilation is seldom used as a life support measure in terminal-stage ALS patients in Europe, in contrast, for example, to some parts of the United States [21]. Indeed, some physicians still argue a priori against mechanical ventilation for ALS patients, without differentiating between non-invasive ventilation via mask (a palliative measure aimed at relieving the symptoms of chronic nocturnal hypoventilation [8]) and tracheostomy, which is intended to prolong the patient's life. One of the arguments cited against mechanical ventilation is the fear of a situation in which the patient might request the physician to discontinue life support. In most European countries, such a request would be denied by many physicians, on the grounds that such an action would be tantamount to "killing upon request", and therefore illegal. Although a shift from the traditional paternalistic attitude (*salus aegroti suprema lex*) to a more patient-centred approach (*voluntas aegroti suprema lex*) is slowly taking place in European medicine, the legal side remains in flux. Since present laws do not cover such cases in many countries, stopping ventilation still entails the theoretical risk of prosecution. However, in the United States, several physicians have already faced criminal prosecution when they *refused* to discontinue ventilation at the patient's request. According to the principles outlined above, this is certainly possible in Europe as well.

The ability to control discontinuation of ventilation is crucial in ALS patients' decisions on whether or not to start ventilation [33]. Therefore, the physician's attitude to this subject must be discussed as part of the patient's information on ventilatory support [28]. It should also be made clear to the patient that consenting to non-invasive ventilation via mask for symptom control does not automatically mean a consent to later tracheostomy. In this context, the role of specific advance directives [24, 30], which need to be periodically revised and updated [28], must be emphasized. If a physician believes that he would not be able to comply with a request by the patient for life support withdrawal, it would seem appropriate for him/her to refer the patient to another doctor for discussion of the ventilatory options.

The cases presented in this article specifically involved the withdrawal of invasive ventilation. It is a more common situation, both in Europe and the United States, that patients who are receiving non-invasive positive-pressure ventilation (NIPPV) wish to discontinue it. In our experience, voluntary termination was one of the most common causes of death in these patients [34]. However, we believe that this situation entails less serious ethical and legal problems, for the following reasons: (1) NIPPV is

never initiated against the patient's will; (2) NIPPV is primarily aimed at relieving the symptoms of chronic hypoventilation, rather than prolonging life [8]; (3) since NIPPV is usually administered at night, the patients can decide every evening whether they want the treatment to be continued; (4) stopping NIPPV does not usually mean immediate death, thus allowing enough time for appropriate palliative measures to be installed. Clearly, some ethical problems still persist, such as the role of the family in a decision that will most likely shorten the patient's life expectancy. The foremost goal should be to ensure that foreseeable symptoms after a treatment-limiting decision (e.g. dyspnoea) are adequately palliated. Hospice institutions can provide invaluable help and assistance to ALS patients in these situations [23]. For patients receiving 24-h NIPPV, the considerations outlined for tracheostomized patients apply.

Since the dichotomy of severe physical debility and unimpaired mental abilities up to the very end is almost unique to ALS, it is not surprising that some of the most spectacular cases of assisted suicide and euthanasia in recent years involved ALS patients [18]. However, the subject of life support withdrawal at the patient's request is a fundamentally different issue [15, 27], in that here a competent patient withdraws his/her consent to an invasive medical procedure. The crucial role of the doctrine of informed consent has traditionally been strongly upheld in the United States [1] and in the United Kingdom [17] and is now gradually becoming accepted as a general principle throughout the European Union.

Obviously, it is the physician's duty to ascertain that the patient's wish is not owing to a reversible condition, such as transient depression. A psychiatric evaluation would appear to be desirable in most cases. It would also fulfill the purpose of having two separate physicians document the patient's wish and mental competence at two different time points.

In our limited experience, we have found that the time point and mode of expression of the patient's wish for life support discontinuation greatly depend on the caring conditions. Therefore, a thorough review of the patient's care situation should be performed prior to any decision about stopping the ventilator. Independently of these circumstances, we believe that it is the ethical obligation of the physician and the caring team to provide the patient with an optimal level of care that stresses the possibility of a fulfilling life despite severe disability. This is exemplified by the subsequent events in case 2: after the physician had reassured the patient that his ventilator would be stopped if he really wished, the patient expressed relief because, as he put it, his worst fear had been to be "trapped" in his ventilator forever. The quality of care was found to be insufficient, mainly owing to a "burn-out" syndrome of the patient's wife and poor external support. Home hospice care was initiated. On subsequent visits, the patient expressed satisfaction with his quality of life and denied any wish for life support withdrawal. He died 6 months later in his sleep, without signs of major distress, possibly owing to untreated aspiration pneumonia.

Conclusions

The main conclusions of this article are:

1. Prevention of unwanted ventilation is a primary goal of ALS patient care. It can be reached by early, frank discussion of the terminal stage with patient and relatives.
2. If a ventilated patient requests disconnection, he/she is not asking to be killed, but is withdrawing his/her consent to an invasive medical procedure.
3. It is the responsibility of the caring team to offer the patient terminal care that provides the chance for a meaningful life even in the presence of severe physical impairment.
4. If, despite these efforts, the patient's will is repeatedly and unequivocally expressed and not owing to a reversible condition such as transient depression, it appears medically, legally and ethically justified for the physician to take the necessary steps, including prior sedation, in order to ensure a peaceful death upon life support withdrawal.

The intricate complex of medical, legal and ethical aspects in cases such as the ones outlined here represents a formidable problem for any single physician [11]. In light of our limited experience, and that reported in the published literature on this subject, we advocate a case-by-case, interdisciplinary team approach, based on (but not slave to) the principle of patient autonomy. The goal is to not unduly restrict the individual patient's freedom of choice in the terminal stage of illness and to ensure that patients are not subjected to invasive medical intervention such as mechanical ventilation once they withdraw their consent to such intervention [6]. However, this is feasible only in a context in which the caring team has done its utmost to provide the patient with a level of palliative care that maximizes the chances for a meaningful life even in the wake of severe physical impairment.

Acknowledgements We thank Prof. U. Büttner, Dr. T. Gasser and Dr. E. A. Oppenheimer for critical review of the manuscript, Dr. J. Bernat for helpful comments, J. Benson for expert editorial assistance and Prof. T. Brandt for support.

References

1. American Academy of Neurology Ethics and Humanities Subcommittee (1993) Position statement: certain aspects of the care and management of profoundly and irreversibly paralyzed patients with retained consciousness and cognition. Neurology 43:222–223
2. American Academy of Neurology Ethics and Humanities Subcommittee (1998) Position statement: assisted suicide, euthanasia, and the neurologist. Neurology 50:596–598
3. Baumann J, Bochnik HJ, Brauneck AE, Calliess RP, Carstensen G, Eser A, Jensen HP, Kaufmann A, Klug U, Koch HG, et al (1986) Alternativentwurf eines Gesetzes über Sterbehilfe. Georg Thieme Verlag, Stuttgart New York
4. Beauchamp TL, Childress JF (1994) Principles of biomedical ethics. Oxford University Press, New York
5. Bernat E (1996) Die rechtlichen Grenzen der Sterbehilfe: gezeigt am Beispiel der amyotrophen Lateralsklerose. Wien Med Wochenschr 146:195–198
6. Bernat JL, Cranford RE, Kittredge FI, Rosenberg RN (1993) Competent patients with advanced states of permanent paralysis have the right to forgo life-sustaining therapy. Neurology 43:224–225
7. Borasio GD, Voltz R (1994) Amyotrophe Lateralsklerose: Fallbericht und medizinischer Kommentar. Z Med Ethik 40:143–147
8. Borasio GD, Voltz R (1997) Palliative care in amyotrophic lateral sclerosis. J Neurol 244 [Suppl 4]:S11–S17
9. Borasio GD, Appel SH, Büttner U (1996) Upper and lower motor neuron disorders. In: Brandt T, Caplan LR, Dichgans J, Diener HC, Kennard C (eds) Neurological disorders: course and treatment. Academic Press, New York London, pp 811–817
10. Brody H, Campbell ML, Faber-Langendoen K, Ogle KS (1997) Withdrawing intensive life-sustaining treatment – recommendations for compassionate clinical management. N Engl J Med 336:652–657
11. Buch AJ (1994) Verantwortete Bewältigung des ethischen Dilemmas. Ethischer Kommentar zum Fallbericht. Z Med Ethik 40:148–153
12. Gaul AL, Wilson SF (1990) Should a ventilator be removed at a patient's request? An ethical analysis. J Neurosci Nurs 22:326–329
13. Goldblatt D (1984) Decisions about life support in amyotrophic lateral sclerosis. Semin Neurol 4:104–110
14. Goldblatt D (1996) Three get ready. Arch Neurol 53:1222–1223
15. Goldblatt D, Greenlaw J (1989) Starting and stopping the ventilator for patients with amyotrophic lateral sclerosis. Neurol Clin 7:789–806
16. Hayashi H, Shuuichi K, Kawada A (1991) Amyotrophic lateral sclerosis patients living beyond respiratory failure. J Neurol Sci 105:73–78
17. House of Lords (1994) Report of the Select Committee on Medical Ethics. HMSO, London
18. Margolick, D (1994) Jurors acquit Dr. Kevorkian in suicide case. New York Times, 3 May 1994:A1
19. McDonald ER, Hillel A, Wiedenfeld SA (1996) Evaluation of the psychological status of ventilatory-supported patients with ALS/MND. Palliat Med 10:35–41
20. Moss AH, Casey P, Stocking CB, Roos RP, Brooks BR, Siegler M (1993) Home ventilation for amyotrophic lateral sclerosis patients: outcomes, costs, and patient, family and physician attitudes. Neurology 43:438–443
21. Moss AH, Oppenheimer EA, Casey P, Cazzolli PA, Roos RP, Stocking CB, Siegler M (1996) Patients with amyotrophic lateral sclerosis receiving mechanical ventilation: advance care planning and outcomes. Chest 110:249–255
22. Müller M, Schöch H (1994) Juristische Stellungnahme zur Sterbehilfe bei amyotropher Lateralsklerose. Z Med Ethik 40:321–326
23. O'Brien T, Kelly M, Saunders C (1992) Motor neuron disease: a hospice perspective. BMJ 304:471–473
24. Olick RS (1996) A disease-specific advance directive for amyotrophic lateral sclerosis patients. Ann Rev Law Ethics 4:553–557
25. Oppenheimer EA (1993) Decision-making in the respiratory care of amyotrophic lateral sclerosis: should home mechanical ventilation be used? Palliat Med 7 [Suppl 4]:49–64
26. Schneiderman LJ, Spragg RG (1988) Ethical decisions in discontinuing mechanical ventilation. N Engl J Med 318:984–988
27. Sherman MS; Paz HL (1994) Review of respiratory care of the patient with amyotrophic lateral sclerosis. Respiration 61:61–67
28. Silverstein MD, Stocking CB, Antel JP, Beckwith J, Roos RP, Siegler M (1991) Amyotrophic lateral sclerosis and life-sustaining therapy: patients' desires for information, participation in decision making, and life-sustaining therapy. Mayo Clin Proc 66:906–913
29. Truog RD, Berde CB, Mitchell C, Grier HE (1992) Barbiturates in the care of the terminally ill. N Engl J Med 327:1678–1682
30. Voltz R, Akabayashi A, Reese C, Ohi G, Sass HM (1998) End-of-life decisions and advance directives in palliative care: a crosscultural survey of patients and health care professionals. J Pain Symptom Manage (in press)
31. Walton DN (1983) Ethics of withdrawal of life-support systems: case studies on decision-making in intensive care. Greenword, Westport, CT
32. Winterholler M, Erbguth F, Rechlin T, Neundörfer B (1997) Der Umgang mit Lebens- und Todeswünschen bei invasiv beatmeten ALS-Patienten. Med Klin 92 [Suppl 1]:90–92
33. Young JM, Marshall CL, Anderson EJ (1994) Amyotrophic lateral sclerosis patients' perspectives on use of mechanical ventilation. Health Soc Work 19:253–260
34. Schlamp V, Karg O, Abel A, Schlotter B, Wasner M, Borasio GD (1998) Nicht-invasive intermittierende Selbstbeatmung (ISB) als Palliativmaßnahme bei amyotropher Lateralsklerose. Nervenarzt (in press)