

Ethical decision-making in two patients with locked-in syndrome on the intensive care unit

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Abstract

Locked-in syndrome (LIS) is one of the most dramatic neurological outcomes and has a profound impact on patients and their families. Most patients have intact cognition and intellectual ability and perception. Communication is possible with eyelid and/or eyeball movement. According to the literature, the wish to die is not an important issue in acute and chronic LIS. This study describes and analyses the ethical decision-making process in two opposite cases of LIS in the intensive care unit. One patient expressed the wish to prolong her life for as long as possible; the other patient asked for deliberate termination of life. Both wishes were honoured. In conclusion, most patients with LIS are competent and intellectually intact. In The Netherlands the autonomy of the patient is respected by law. In respecting this autonomy, medical choices can be different in comparable patients.

Introduction

Although locked-in syndrome (LIS) is rare,^{1,2} it has become better known to the lay public since the 2007 release of Julian Schnabel's film version of the book *Le scaphandre et le papillon* ('The diving bell and the butterfly'). The book was 'written' by the 43-year-old LIS patient Jean-Dominique Bauby, by blinking his left eye in response to reading an alphabet. A friend recited the letters and recorded the choices, forming the words and sentences of the resulting book.^{3,4}

The incidence of LIS is difficult to determine from the medical literature, largely because it is often misdiagnosed as coma or vegetative state. Long-term survival in LIS is rare, and death occurs within the first four months in 87% of cases.⁵ However, chronic LIS patients report a meaningful quality of life and their demand for euthanasia is rare.⁶ In 1993 the American Academy of Neurology published a position paper concerning the care of conscious, competent patients with profound paralysis.⁷ They concluded that such patients have the right to make health-care decisions about their own lives, including

acceptance or refusal of life-sustaining treatment. The following quotation is from the position paper:⁷

'Once patients have decided to forego life-sustaining treatment, physicians have an ethical obligation to minimize their subsequent suffering. This is particularly true of profoundly paralyzed patients, because cognition and sensation may be intact, and they are capable of great suffering.'

According to the literature, the wish to die is not an important issue in acute and chronic LIS.^{6,8} One physician found it appropriate to request psychiatric consultation for competency determination in the case of a young woman with a traumatic quadriplegia who requested a do-not-resuscitate (DNR) order.⁹ The psychiatric consultant declared her to be incapable of medical decisions because depressive feelings had impaired her judgement.

In the present article, the ethical decision-making in two cases of LIS patients in the intensive care unit (ICU) is described.

Case 1

A 45-year-old woman was admitted with a one-hour history of nausea, vomiting and vertigo. Her condition had deteriorated within a few hours; she complained of severe vertigo and slurred speech, and right-sided hemiparesis was observed. A computed tomography (CT) scan of the brain showed an area of increased density in the pons, ventral to the fourth ventricle, resulting from an occlusion of the basilar artery. She was transferred to our hospital for further treatment.

The patient was sedated with propofol, intubated and ventilated with continuous positive airway pressure

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(CPAP). After ICU admission, the sedation was stopped for neurological examination. The next day she could withdraw her right leg in response to painful stimuli, but showed no movement of her arms or left leg. In the evening of the same day, she opened her eyes with a positive vertical gaze, but was unable to move her eyes horizontally. A left-beating nystagmus was observed. Thrombolysis was started a day after the insult. That night she showed a flaccid tetraplegia, a vertical left-beating nystagmus and vertical eyeball movement on request, but horizontal eye movement was impossible. Movements of the jaws, tongue and head were absent. She was drowsy, but her reactions were adequate. Her vision and hearing seemed to be unchanged. She was diagnosed with LIS based on the basilar artery occlusion.

On the fourth day, the patient, her husband and two adult daughters were informed about the diagnosis and prognosis. Two possible plans were offered: removal of the endotracheal tube and sedation leading to the death of the patient, or removal of the endotracheal tube with subsequent tracheotomy and further treatment. The patient was given some time to think this over. The next day the two possible scenarios were offered again and in addition the possibility of deliberate termination of life (euthanasia). The patient communicated by eye blinking that she wished prolonged treatment. She was aware that motor recovery was very unlikely. Her husband communicated to the intensivist that he wished withdrawal of treatment, but that he respected the wishes of his wife to prolong her life as much as possible.

Some of the health-care professionals had second thoughts about the competence of the patient to make adequate decisions and wanted to discuss what to do in the case of complications or whether chronic mechanical ventilation could be offered to this patient. An *ad hoc* ethical deliberation was planned for the same afternoon. Two intensivists, a neurology fellow, an ICU fellow, a clinical ethicist specializing in intensive care and 12 ICU nurses were present during this deliberation. Prior to the discussion, several facts about LIS, extracted from recent publications, were presented:

- (1) Most patients with LIS cases of pure brainstem lesions have intact cognition and intellectual ability and perception. Communication is possible with eyelid and/or eyeball movement.^{10,11} Some patients have cognitive impairment in the form of slower cognitive processing speed and perception;^{12,13}
- (2) The mortality of LIS resulting from vascular pathology is high (76%) where the majority of patients (87%) die in the first four months after the primary insult;⁶
- (3) Almost all patients require a tracheotomy in the acute phase, but 75% of those who survive can breathe spontaneously in the chronic phase;⁵
- (4) All patients with LIS are totally dependent on care for the rest of their lives, which can give rise to a long-term physical and emotional burden on both the patient and the patient's family;
- (5) Despite the fact that most patients remain severely disabled, only a few want to die;⁶

- (6) In The Netherlands, competent patients with acute LIS can 'ask' for deliberate termination of life, which is a request that can be honoured.¹⁴

In the ensuing discussion, consensus was reached on the ethical, judicial and practical consequences of the intact cognition and competency of the patient. Based on the Dutch Medical Treatment Contracts Act (1992),¹⁵ a competent patient has the right to choose prolongation of life if this is technically possible and if the physicians think it is reasonable to do. The patient can, under the same Act, also ask for withdrawal of treatment or a DNR order. Tracheotomy and gastroenterostomy are considered appropriate care by the physicians and nurses; however, in cases of severe complications (cardiac arrest, cerebral haemorrhage or severe sepsis), further treatment should be withheld due to the high risk of secondary cerebral damage. Prolonged mechanical ventilation outside the hospital is considered to represent adequate care. Deliberate termination of life, requested by a competent LIS patient, was judged to be an acceptable choice that should be honoured.

On the ninth day, a tracheotomy and subcutaneous gastroenterostomy were performed. On the nineteenth day, the patient was weaned from the mechanical ventilation and was breathing adequately. She was discharged from the ICU to the stroke unit of the Department of Neurology. A rehabilitation programme was initiated. However, on day 31 she unexpectedly lost consciousness due to an acute haemorrhage in the brainstem. The patient was deeply comatose, but had spontaneous regular breathing. There were no treatment possibilities and she was allowed to die. Later that day, a Cheyne-Stokes breathing pattern was observed. Shortly afterwards, she died without any signs of discomfort, in the presence of her relatives.

Case 2

The second case has previously been described in detail,¹⁴ and is presented again here as an illustrative contrast to the first case. A 56-year-old man was admitted to the ICU after developing an unresponsive state. Upon examination the patient was found to be unconscious and tetraplegic. A CT scan showed a bilateral haemorrhage in the pons. Due to respiratory deterioration, the patient was intubated and mechanically ventilated.

The patient's family described him as an intelligent, independent and previously active man. No written advance directive concerning his wishes regarding life-sustaining therapies existed, but his spouse stated emphatically that he would reject continued therapy in the case of being in a totally dependent state. His wife characterized him as a proponent of euthanasia, and asked the physician about the possibility of euthanasia. The physician had no personal objections in general, but indicated that the comatose patient was not suffering, and that there was no written request from him. On the sixth day, the patient opened his eyes on request. Hearing, vision and bodily sensation were normal. The diagnosis of LIS resulting from a pontine haemorrhage was made.

In the second week, the patient learned to communicate by eye opening, and was informed about his diagnosis and prognosis. On the 12th day, he was asked if he understood his diagnosis and prognosis. He indicated 'yes'. Asked whether he was in pain, he indicated 'no'. Again, he was told that survival could be prolonged for years with adequate care and mechanical ventilation. He was asked whether he wished such treatment. He indicated 'no'. Did he understand that he would die without such continued care? He indicated 'yes'. Did he want the invasive treatment to be withdrawn? He indicated 'yes'. His relatives confirmed that this was absolutely in line with his character. The intensivist asked the patient whether he desired death due to withdrawal of mechanical ventilation and subsequent administration of sedatives. He indicated 'yes'. He was told that he was not dependent on mechanical ventilation (CPAP) and that he would be able to breath under sedation for hours or days during which time there would be respiratory deterioration and exhaustion before death. Another possibility was deliberate termination of life. On the question of whether he would like to end his life by injection of life-terminating agents, he also indicated 'yes'. The physician documented these thoughts in the medical chart and asked an intensivist from another ICU of the same hospital for a second opinion. Questions to be answered included: 'Is the patient competent?', 'Is the [indirect] request for euthanasia based on valid grounds and is the patient's suffering lasting and unbearable?', and 'Is the patient informed about his situation, is the request well considered, and do any reasonable alternatives to euthanasia exist?'

The second intensivist visited the patient and his relatives on the eighteenth day. He concluded that the diagnosis was valid, that the condition was fatal without invasive treatment and 24-hour care, that the patient's view on life and death was well-considered (also based on an intensive conversation with the relatives). He reconfirmed that all provisions for euthanasia were fulfilled. Since this represented a unique situation, a third physician was also consulted. His conclusion was in agreement with those of the other physicians. The board of the hospital was informed about the intended euthanasia. On the 26th day, the patient was again asked if he still wished for deliberate termination of his life; the patient confirmed the request. The intensivist discussed the situation with the nursing staff, who had no professional objections. Euthanasia was scheduled for day 33. The patient was breathing spontaneously on CPAP only. On day 32 friends visited the patient for the last time. On the following day a senior ICU nurse and the intensivist prepared the euthanizing agents: 30 mg midazolam, 1.4 g pentothal (20 mg/kg) and 20 mg pancuronium. Close relatives were present at the bedside. After everyone had said farewell to the patient, the intensivist injected 30 mg midazolam intravenously, after which the patient fell peacefully and quietly into a deep sleep. The ventilator was disconnected and the intensivist subsequently intravenously injected 1.4 g pentothal. Within five minutes, ventilatory arrest was observed and subsequent circulatory arrest and deep

arterial hypotension were visible on the monitoring equipment. Ten minutes later, the patient was declared dead. There was no need for injection of the muscle relaxant pancuronium. The regional review committee reviewed the case and questioned the physician, but concluded that the physician had acted within the requirements of due care.

Discussion

LIS represents an extreme form of bodily imprisonment.¹⁶ However, intellectual capacity is usually preserved, though some will claim that patients have an impaired capacity to make valid decisions about their health-care status, especially in the acute phase of the condition.^{9,13,17} Crucial in the decision-making process in the two cases reported was respect for the autonomy of the patient. Based on Dutch legislation, health-care professionals have to respect the wishes of the patient if these are understandable and within the accepted possibilities of medical care and also if the patient is judged to be competent to make these judgements. In Case 1, after holding an ethical discussion, health-care professionals had no reasons to doubt the cognitive functioning and competency of the patient. For these reasons, her preference for continuation of treatment was followed. However, if certain aspects of the treatment are judged to be futile, physicians have no obligation to continue treatment, even if the patient asks for it. In the first case, resuscitation in the case of circulatory arrest in treatment for severe sepsis was judged to be futile, so these procedures must be withheld. In contrast to the case description of Field,⁹ we would comply with a DNR request of a patient with LIS or quadriplegia, respecting her autonomy and in accordance with Dutch law.

The second case is far more unique and complicated, and many will judge the decision to be inappropriate, or even immoral. The Netherlands was the first country in the world to enact a law on euthanasia. Euthanasia is defined as the intentional termination of life of an adult patient upon request. This presupposes voluntariness of the patient and a deliberate active action by the physician. This excludes every form of intentional involuntary termination of life (as active termination of life in comatose patients or neonates). In the Dutch Termination of Life on Request and Assisted Suicide (Review Procedures) Act (2002),¹⁸ the requirements of due care are described. The physician

- (1) Holds the conviction that the request of the patient is voluntary and well-considered. Holds the conviction that the patient's suffering is lasting and unbearable. Has informed the patient about the situation and prospects;
- (2) Holds the conviction that there is no other reasonable alternative in light of the patient's situation;
- (3) Has consulted at least one other independent physician who must have seen the patient and given a written opinion on the due care criteria referred to above;

- (4) Has terminated a patient's life with due medical care and attention.

One of the foundations of the Euthanasia Act is, as in the Medical Treatments Contracts Act, to respect the individuality of the patient. Patients with LIS are unable to speak or write, so health-care professionals can only ask the patient to express his or her will on treatment options, as we did in both of the described cases. In our second case, there were no doubts about the competence of the patient, and after several conversations with the close relatives of the patient, we had a clear picture of the patient's character and way of life, which made his request for life termination understandable. The dominant non-physical suffering would be lasting. From the physician's perspective, euthanasia is based on the ethical principle of respecting the patient's autonomy and on mercy. The duty to alleviate unbearable and lasting suffering has, via the justification of necessity, become the basis for euthanasia. We believe that it is inhuman to keep a conscious tetraplegic patient alive against his will. Nick Grisholm, who had lived with LIS since 2000, stated: 'I have thought of suicide often. Even if I wanted to do it, now I could not, it's physically impossible'.¹⁹ Several authors have stated that LIS patients have no wish to die; yet the question remains: is it that the patients do not want to die, or is it that physicians do not offer the possibility of treatment withdrawal? We think it is disrespectful to the patient and paternalistic to neglect a patient's wish to die. The second case is unique ethically because deliberate termination of life on request is, even in The Netherlands, rare when it is performed within a month after the diagnosis, and in many countries is ethically and legally in conflict with the moral principle of respect for life. It is clinically unique because the patient communicated his request for euthanasia without speaking or writing, but only by blinking his eyes.

In conclusion, most patients with LIS are competent and intellectually intact. As Emile Zola described in his novel, *Thérèse Raquin*, it is like being 'buried alive in a dead body'.²⁰ The patient is locked inside his body. It should be the choice of the patient if he or she wants to continue such a life. With the Dutch Medical Contract Act and Euthanasia act, we think that we can provide our LIS patients with care in accordance with their view on life, death and dignity.

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The ethics committee of the Erasmus MC University Medical Center Rotterdam, The Netherlands, approved

publication of Case 2. The husband of the patient in Case 1 gave his consent for publication.

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