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BIOETHICAL ISSUES IN THE ORGANIZATION OF SPECIALIZED MEDICAL CARE FOR PATIENTS WITH MOTOR NEURON DISEASE IN THE REPUBLIC OF SAKHA (YAKUTIA)

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Amyotrophic lateral sclerosis (ALS) is a severe neurodegenerative disease in which the death of central and peripheral motor neurons occurs, leading to an inevitable fatal outcome. Bioethical problems arise at any stage of the disease when a patient with ALS goes to a doctor. The help of psychologists, the activities of bioethical committees in medical organizations in informing patients and their relatives about the disease, the awareness of neurologists about social support measures from the state can help in organizing medical and social assistance measures to improve the quality of life of patients with ALS.

Aim: to solve the bioethical problems of patients with ALS to improve the organization of specialized medical care

Keywords: bioethics, motor neuron diseases, amyotrophic lateral sclerosis, artificial lung ventilation, palliative care.

Introduction. ALS is a serious disease of unknown etiology and pathogenesis, accompanied by the death of central and peripheral motor neurons, steady progression and inevitable death. The incidence of ALS in the world is 1.89 per 100 thousand population, and the prevalence is 5.2 cases per 100 thousand population [17]. 7% of ALS patients have been ill for more than 5 years, the average life expectancy is 2.5 years with bulbar and 3.5 years with spinal debut of ALS. In recent years, there has been an increase in the incidence of MND in the world [11].

The disease is manifested by the development of motor disorders, bulbar and pseudobulbar syndrome, as a result of which the patient becomes immobilized, cannot eat and talk. The main cause of death in patients with ALS is restrictive or restrictive-obstructive respiratory failure, which develops due to paralysis of the diaphragm muscles, respiratory muscles and aspiration of food and saliva in bulbar disorders[7].

Managing a patient with ALS is a very difficult task for a doctor because of the severity, rapid progression and incurable course of the disease. In a challenging complex of medical and social objectives, one of the main ones are the bioethical problems that accompany a patient with ALS until the end of his life. The complexity of bioethical problems arises at all stages of the provision of specialized medical care (SMC) by a neurologist. At the 1st stage of outpatient SMC, if ALS is suspected, the doctor is faced with a dilemma: whether to inform the patient and his relatives about his suspicions or await the final diagnosis after the examination. When providing SMC at stage 2 in an inpatient hospital, when differential diagnosis with ALS-like diseases has been carried out and the final diagnosis of ALS has been established, the attending physician also faces the difficult task of communicating the diagnosis to the patient and his relatives. At the same time, psychological work should be carried out separately with the patient and his relatives who will take care of him in the future. The decision on palliative therapy, social, financial issues, as well as the patient's preliminary orders should be made long before there is a need for enteral nutrition or auxiliary ventilation of the lungs. All this requires the use of a complex of social, medical, and legal measures, the main of which is a bioethical issue affecting the moral aspects both on the part of the doctor and on the part of the patient and his relatives. In the Russian Federation, there is no single standard for the management of patients with incurable and fatal diseases, including ALS.

Materials and methods of research. The study included patients with ALS (n=11), their relatives (n=11) and neurologists (n=30). 3 out of 11 patients needed periodic artificial lung ventilation. All study participants gave written informed consent to participate in the study.

Inclusion criteria:

1. patients with clinically reliable ALS using El Escorial criteria [5];
2. relatives of ALS patients who do not have a mental illness;
3. doctors with a specialization in neurology

Exclusion criteria:

1. patients with ALS-mimicking syndromes;
2. patients and their relatives who have not signed a written informed consent for the study;
3. patients with severe MND who cannot participate in the study on their own;
4. relatives of patients with mental illnesses.

Research methods

1. The clinical method included a study on El Escorial criteria to establish ALS and to assess somatic status
2. Using the Hospital Anxiety and Depression Scale (HADS) to study the psycho-emotional state of patients and their close relatives.
3. The method of medical interview was conducted with patients and close relatives caring for a patient with ALS to determine the degree of readiness to accept information about their disease.
4. The questionnaire method was conducted for neurologists.
5. Statistical analysis of the research results was carried out using Excel to determine the average values.

Results. The sample of patients included 11 patients with reliable ALS according to El Escorial criteria. By ethnicity, 8 (72.7%) people are representatives of the Yakut ethnic group, 3 (27.3%) pa-

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tients are of the Russian ethnic group. The average age was 53±11.8 years. The age varied from 30 to 72 years. In 5 patients, the diagnosis of "ALS" was established for the first time. The remaining 6 patients were diagnosed from 1.5 to 5 years ago.

A survey on the HADS scale showed that all patients experience clinically pronounced anxiety and depression. However, a survey on the HADS scale of relatives of patients did not reveal clinical signs of depression, while the level of anxiety also significantly exceeded normal indicators. Thus, in the group of patients with ALS, the anxiety level averaged 17.74± 0.48 points, depression - 13.60± 0.68. In relatives of patients with ALS, the anxiety level was 13.8 ± 0.32 points. The average level of depression in family members was 5.9±0.62 points.

Thus, our study shows that all patients and their family members experience pronounced psycho - emotional stress associated with the disease.

In addition, in none of the cases was the help of a clinical psychologist provided, because in outpatient clinics to which these patients were attached, the staffing of this specialist is not provided due to the absence of a compulsory medical insurance (CMI) tariff for this medical service. In inpatient hospitals, excluding hospitals of the regional vascular center, vacancies of clinical psychologists are also not provided for this exact reason. If possible, this function is performed by the attending neurologist.

During the survey , neurologists answered the following questions:

1. Do you think that it is necessary to hide the diagnosis under an ALS-like disease, for example, vertebrogenic myelopathy, so as not to cause depression, suicide, vascular crises in the patient, which can lead to acute disorders of cerebral circulation and acute coronary syndrome, etc., or give them full information about the disease?. 7.4% of doctors answered affirmatively, and 92.6% of the surveyed doctors believe that it is not necessary to conceal the disease from the patient;

2. Do patients with ALS and their relatives require the help of a psychologist? 100% of the respondents answered "Yes".

3. Does your organization have a psychologist?

A survey among doctors of state budgetary and autonomous institutions showed that 48% of these institutions have a psychologist;

4. Does your organization have a local bioethics committee?

The survey showed that only 20% of medical organizations have local bioethics committees;

5. Are you familiar with the contents of the Order of the Ministry of Health of the Russian Federation No. 348n "On approval of the list of medical devices intended to maintain the functions of organs and systems of the human body provided at home" dated May 31, 2019, the list of which includes general-purpose artificial lung ventilation devices provided for use at home. 19% of doctors were not familiar with this order;

6. What measures of medical and social assistance would you recommend?

Neurologists recommend these measures of medical and social assistance:

- supervision by multidisciplinary teams, including home visits – 3.2% of the surveyed doctors;
- management of patients by specially trained neurologists and treatment of patients in specialized ALS clinics– 31,2%;
- creation of medical and social care centers for patients with ALS – 3.2%;
- creation of a unified ALS register – 3.1 %;
- creation of local bioethics committees – 51.1%.
- simplification of the procedure for obtaining at-home-ventilators through Palliative care centers – 11.4%

The method of clinical interview revealed that patients were concerned about the following questions (each question out of 100% of cases):

1. How will ALS affect life expectancy - (92%)?
2. Will they be able to cope with their daily work – (85%)?
3. Is this disease inherited – (65%)?
4. Why did the patient get this disease – (97%)?
5. Is there a search for an effective drug – (86%)?
6. Are there state support measures for patients with ALS (45%)?

Discussion. Observation of patients with ALS revealed bioethical problems at different stages of disease progression. The patient's condition depends on how the diagnosis is presented: will the patient be able to accept the inevitability of a fatal outcome, will he choose a position of humility and non-resistance to the disease or will he find the inner strength to live with this incurable and fatal disease. And this circumstance imposes on the attending physician a great moral responsibility in connection with the need to communicate the diagnosis to the patient and people close to him. When communicating the diagnosis of ALS to the patient and his loved ones, it is necessary to observe the moral norms of medical ethics and deontology.

Bioethical problems in reporting the diagnosis of amyotrophic lateral sclerosis. Guided by paragraph 1 of Article 22, Federal Law No. 323 of 21.11.2011 (ed. of 03.07.2016), the doctor must provide all the necessary information without the intent of concealment. In case of an unfavorable prognosis, the nature of the disease should be reported in a delicate form to a citizen or his spouse, one of the close relatives, unless the patient has forbidden to inform them about it and (or) has not identified another person to whom such information should be disclosed." Due to the lack of a single algorithm of actions in the Russian Federation in such situations, the doctor acts at his discretion and in the way it is customary to act in these cases in this medical institution [1]. When deciding on the diagnosis of "ALS", it is necessary to keep in mind how the patient and his relatives will perceive the diagnosis.

When voicing the diagnosis, the attending physician needs to take into account the characteristics of the patient's character, how close his ties are with family members, whether there are other close people besides relatives. It is important to assess the situation in advance and choose the tactics of voicing the diagnosis: will the patient be able to accept the diagnosis alone or is it preferable to communicate the diagnosis in a circle of relatives or close people. The support of relatives and close friends during the period of informing about a fatal disease is very important[9].

Thus, the doctor must foresee what emotions the news of his diagnosis will cause in the patient and be internally prepared for them in order to be able to react objectively. Sometimes doctors, in order to avoid a difficult moral situation when communicating with a patient, postpone the diagnosis until the day of discharge from the hospital. Such a position on the part of a doctor, on whom patients pin their hopes, is undesirable and unacceptable. We believe that the diagnosis should be reported before discharge from the hospital so that the patient can collect his thoughts and assess the situation with his relatives, as well as discuss what medical and social assistance he can receive after discharge from the hospital.

Bioethical problems when connecting the patient with ALS to a ventilator. For the palliative treatment of ALS, there is a "gold standard" of palliative therapy for ALS, which includes artificial lung ventilation, percutaneous endoscopic gastrostomy, enteral nutrition, the drug riluzol

(not registered in the Russian Federation) and symptomatic pharmaceutical and non-pharmaceutical treatment [8].

Medical personnel should remember that the installation of a ventilator to replace respiratory function requires prior resolution of bioethical problems. Before installing the device, it is necessary to have a conversation with relatives and the patient. It should be noted that the decision to connect a patient to a ventilator primarily falls on his family and is a very important moral step on their part, since the maintenance of the life of a sick family member depends on the care of a patient with a ventilator. In some cases, the patient decides not to continue communicating with doctors and refuses to discuss his diagnosis with loved ones, falls into despondency and refuses to be treated [15]. In such cases, not only the help of close people is needed, but also a psychologist, so that they can help the patient cope with the awareness of the disease and direct him to the path of solving the problem, and not escaping it. The sources describe cases of affective behavior and numerous suicidal attempts in such situations, for example, in Huntington's disease [9]. When choosing to connect to a ventilator, the patient's family or close circle should take into account their financial capabilities, familiarize themselves with the requirements for caring for a patient who is on a ventilator in a hospital or at home. Before connecting the ventilator, it is necessary to obtain informed consent for the installation of the device.

Using a ventilator at home, until June 2019, was an unsolvable problem for most patients with ALS, not only because of the financial difficulties of purchasing equipment for personal use at home, but also the lack of state support. On May 31, 2019, two Orders were issued by the Ministry of Health of the Russian Federation, the Ministry of Labor and Social Protection of the Russian Federation No. 348n "On approval of the list of medical devices intended to maintain the functions of organs and systems of the human body provided at home" [3] and Order No. 345n/372n on approval of the "Regulations on the organization of palliative care, including the procedure for interaction of medical organizations, social service organizations and public associations, other non-profit organizations engaged in their activities in the field of health protection" [4]. In Order No. 348n, the list includes general-purpose artificial lung ventilation devices provided for use at home. These orders are a good state support for patients receiving palliative care at home, about which patients and medical workers are insufficiently informed. Out of all the surveyed neurologists, 88% indicated insufficient provision of ventilators.

Around the world, an interdisciplinary approach to the symptomatic treatment and care of patients with ALS is practiced, which could be adopted and implemented in Russian healthcare at the level of primary medical and social assistance [10, 12, 16].

The preservation of intelligence of patients with ALS, on the one hand, helps them to objectively assess their current situation and independently or jointly with loved ones - solve the issue of gastrostomy and periodically connecting to a ventilator, and on the other hand, they understand and are aware of the fact that palliative care is only a temporary measure. At the same time, the absence of bedsores, replacement respiratory therapy with a ventilator due to good care, as well as the participation of the MND patient's loved ones in solving his issues, make the diagnosis of amyotrophic lateral sclerosis not so fatal. An interdisciplinary, palliative approach can prolong survival and preserve the quality of life [10,13,14]. This requires clarification to relatives that with timely prevention of complications, i.e. adequate patient care, proper ventilation, a patient with ALS can live on a ventilator for an average of up to 1 year or more [2].

In our study, two patients with ALS underwent respiratory replacement therapy with a ventilator at home. This is a woman, 54 years old, Sakha, with a bulbar debut of ALS and a moderate rate of progression of the disease and a man, 54 years old, Russian, with a cervical debut of ALS and a rapid rate of progression. In the first case, before connecting to the ventilator, a preliminary conversation was held with the patient and her husband, informed consent was obtained to connect her to the ventilator at home. Before purchasing a ventilator, the patient was in the intensive care unit. The ventilator was purchased at their own expense. The woman lived for 5 years after connecting the ventilator, nutrition was carried out through a nasogastric probe. The patient was periodically examined by a neurologist, therapist, surgeon, received physical therapy, massage. That is, a multidisciplinary approach was implemented during her care. In the second case, the life expectancy with a ventilator was 2 years. Given the rapid rate of development of the disease, it was decided to inform his wife about the possible imminent death of a patient with ALS, who asked not to inform her husband about it, because the patient himself was in a state of subclinical depression. Since the FVC was less than 50%, the patient had indications for connection to a ventilator. The couple decided to connect to a ventilator. After receiving written informed consent, a ventilator was installed for the patient. In the future, a ventilator for use at home was rented.

Doctors and relatives of the patient should take into account the inevitable emotional and volitional disorders, as well as depression in ALS, which can lead to untimely decisions by the patient regarding treatment methods aimed at prolonging life. It is necessary to convince the patient that the use of a ventilator at home is a method of respiratory support, and not a resuscitation measure, and does not require the patient to stay in the intensive care unit. At the same time, the patient has the opportunity to stay with his family, can travel and even work remotely. Family members should be warned about the reorganization of their everyday life, adjusted for patient care. All patients and their family members need the help of a psychologist and a psychotherapist.

Conclusion. Thus, our study of bioethical issues in the organization of specialized care for patients with amyotrophic lateral sclerosis revealed both the insufficiency of existing measures of medical and social support, and the insufficiency of informing doctors about regulatory documents of state support for patients who need a ventilator, the absence of psychologists and bioethical committees. Psychological support during the entire period of observation of patients, compliance with ethics and deontology, respect for the rights of the patient are the basis for the organization of specialized care for this category of patients. Local ethical committees of medical institutions should help in discussing complex ethical problems that have arisen in clinical practice and need to formulate recommendations on how these problems should be solved. It is also necessary to introduce the experience of interdisciplinary teams in the management of patients with ALS.

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