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DOI 10.25789/YMJ.2024.85.23

UDC 614.253:17.023

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BIOETHICAL PRINCIPLES AND APPLIED BIOETHICS IN THE FIELD OF NEURODEGENERATIVE DISEASE RESEARCH IN YAKUTIA

Applied bioethics is a field of knowledge, the subject of which is practical moral problems. The ethical principles that form the basis for applied bioethical research related to neurodegenerative disease in Yakutia are discussed. Applying the basic four ethical principles as starting points can lead to different answers regarding specific bioethical problems, in our case the moral problems of providing medical care to patients with neurodegenerative diseases.

Keywords: bioethics, applied ethics, neurodegenerative diseases, Republic of Sakha (Yakutia)

Introduction. Bioethics arose from the need to assess the positive and negative effects of new methods and technologies. Can bioethics prevent the unfair distribution of medical resources, treatments, and medicines among those in need? Bioethics acts as a kind of discussion platform, where scientists themselves, who have created breakthrough technologies, discuss the positive and negative consequences of inventions for both patients and society, and positive and negative criteria depend on knowledge, cultural level and prejudices of the era and society as a whole [8,9,11,46].

Perhaps it is bioethics that is the field of human knowledge where many areas of research have become entrenched and there are many directions for research: the philosophical study of the ethics of medicine, medical law, medical anthropology, medical genetics, medical sociology, health policy, health economics. Philosophers believe that bioethics is one of the branches of practical (or applied) ethics, which, in turn, is one of the branches of philosophy [5,12,45].

On the other hand, there is undoubtedly a problem that the philosopher Benatar (2006) calls the problem of "disciplinary slip", when a person moves from working in his own discipline, in which he is trained, to working in another, in which he is not trained. There are fewer obstacles for health care workers or scientists to engage in moral philosophy than there are for philosophers who would like to practice medicine. This does not prevent lawyers, doctors and scientists of various profiles from cooperating with philosophers on practical ethical issues or working independently to answer questions that are crucial for making ethical decisions [22,24,26,43].

According to Husseinov (2004), ethics is initially a practical science. In his opinion, ethics is a practical science, since it considers human actions and behavior from the point of view of their fundamental principles, and it is the point at which philosophy connects with practice, there-

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fore it is rightly called practical philosophy [6]. Thus, applied bioethics is a field of knowledge and behavior, the subject of which is practical moral problems [6, 23,29,45].

Tom Beauchamp and James Childress in 1977, in the book Principles of Biomedical Ethics, proposed standard approaches to bioethics, which formed the basis for the principles of health ethics, along with the Hippocratic Oath. Gillon R (1994) develops an approach that is easy to comprehend ethical problems - "four principles and the scope of their application." It is based on four common, basic prima facie moral obligations - respect for autonomy, charity, nonviolence and justice, plus concern for the scope of their application, i.e. a common, basic moral and analytical framework and a common language are proposed. Although they do not contain orderly rules, these principles can help doctors and other medical professionals make decisions by reflecting on the moral issues that arise at work. Thus, four ethical principles are proposed: "do no harm" (non-maleficence), "do good" (beneficence); the principle of respect for personal autonomy; justice and four rules: veracity, privacy, confidentiality, fidelity [25,28,31,32,47].

In modern society, with the rapid development of technology, it is quite problematic to find ideal solutions to bioethical problems. "It cannot be said that all decisions have been made. On some issues, an ideal compromise may never be found. But most importantly, the discussion continues," M. Vorontsova emphasized (2024) following the results of the online conference "Bioethics and Genetics: challenges of the XXI century" [4,5].

The purpose of this review is to discuss four ethical principles that are the basis for our applied bioethical research related to the problems of neurodegenerative diseases (NDD) in Yakutia. The provision of medical care to patients with NDDs is associated with many moral issues that require close attention from medical professionals and society. The article outlines bioethical principles and provides examples from practical work, that is, certain results on applied bioethics related to our desire to observe the moral principles of medicine.

Principles of bioethics. *The principle of "do no harm" (non-maleficence).* This principle is fundamental in the ethics of medicine. The moral duty of a Hippocratic physician is to provide the most complete medical care to the patient without causing him any physical or psychological damage. However, a moral conflict arises immediately. As soon as the doctor begins his manipulations, he inevitably risks causing pain and moral suffering to the patient. According to Yudin (1998), the harm coming from a doctor should only be objectively inevitable and minimal harm. In addition, the harm that a doctor's actions can bring to a patient can be intentional or unintentional. Intentional harm occurs in cases of inaction by a doctor in a certain situation, or intentional harm. In cases where the doctor did not have the opportunity to think about the possible consequences of the intervention or uncontrolled external influences arose, we can talk about unintentional harm [21].

Gillon R (1994) emphasizes that a physician should provide a net medical benefit to patients with minimal harm, that is, benefit without harm to health. To achieve these moral goals, health care workers have a wide range of obvious obligations. What are the main obligations of medical professionals? Firstly, effective and continuous training both before and during professional activity; secondly, obtaining valuable empirical information during treatment and as a result of medical research and using it to minimize risks to the patient; thirdly, empowering the patient/client with respect for their autonomy to achieve a common goal [31,32,37].

The principle of "do no harm" is the central ethical issue of our biomedical activities in providing medical care to patients with NDDs and their relatives.

We started bioethical research simultaneously with the introduction of DNA diagnostics of spinocerebellar ataxia type 1 (SCA1) into the clinical practice of medical and genetic consultation at the Republican Hospital No. 1 of the National Center of Medicine. Due to the late-manifesting nature of the hereditary disease, seeking the advice of a patient with the risk of carrying the SCA 1 mutation is accompanied by great moral and psychological stress, under the influence of strong emotions, a person may experience doubts and fear. The task of the doctor is to align ethical principles with the different expectations of his patients and help them adapt to the decision on the need for examination and molecular genetic diagnosis. Following the principle of "do no harm", we adhere to the ethical rule of "non-directivity and voluntariness" in consulting.

The most difficult bioethical problem in our practice is presymtomatic DNA testing. Obtaining a positive result of DNA diagnostics, indicating the detection of an SCA 1 mutation, is a "sentence" for the patient, the psychological consequences of which undoubtedly affect the further life of the individual. Therefore, the ethical rules we have adopted provide for minimizing the damage from negative information, namely, respecting the patient's choice in his desire to conduct a DNA diagnosis, or to abandon it.

The principle of "do good" (beneficence). The principle of charity refers to the moral and psychological component of medical care. A person who has chosen the profession of a doctor undertakes to be spiritual, compassionate, positive, and merciful. According to Gusseynov (2004), medical ethics is aimed at transferring the truth and the mystery of morality, the highest meaning contained in it, to medical practice. Medical activity rises to the level of mission, service; it is not just that a doctor is required to be selfless and other moral qualities, but his very activity is given a moral status, as if it were not just a professional job, but also a kind of sacrament. It is considered morally valuable in itself. Medical ethics proceeds from the presumption that adequate behavior within the framework of medical activity cannot but be moral [6]. Moral norms are not fixed by laws and, for this reason, can be interpreted by each person arbitrarily, at his discretion, according to his morality. Their implementation is controlled not by laws, not by coercion, but by spiritual influence, public opinion [20].

Thus, the central question regarding charity in the patient-doctor relationship is: “What does it mean for a doctor to strive for a greater balance of benefit and harm in patient care?” The charity model answers this question, at least from the point of view that medicine is based on the best interests of the patient, not the doctor. The model clearly explains that the central theme of charity is the duty of the doctor to benefit patients, that is, charity as a principle of medical ethics asserts the obligation (on the part of the doctor) to help others (patients) in the realization of their important and legitimate interests and to refrain from harming them in any way, that is, psychologically, morally or physically. In addition, the achievement of the common goal of well-being is carried out by the joint efforts of the doctor and the patient [25,37,39].

The joint work of charitable organizations and state executive bodies, in particular the healthcare system, can help solve the ethical problems of maintaining life in incurable patients with motor neuron diseases (MND). The issue of using an artificial lung ventilation device (ventilator) at home for most patients with amyotrophic lateral sclerosis (ALS) is a big problem mainly due to the difficulties of purchasing ventilator equipment for personal use at home. On May 31, 2019, orders of 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" [15] and Order No. 345n/372n on approval of the "Regulations on organizations providing palliative care, including the procedure for interaction between medical organizations, social service organizations and public associations, and other non-profit organizations engaged in their activities in the field of health protection" [16]. Order No. 348n includes general-purpose ventilators provided for use at home.

It is necessary to convince the patient that the use of a ventilator at home is a method of respiratory support, not an intensive care measure, and does not require the patient to stay in the intensive care unit and anesthesiology. 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 to care for the patient. All patients and their family members need the help of a psychologist and a psychotherapist [2.13].

The principle of "do good" is also implemented by the work of the charity project "Green Cane" within the framework of the activities of the Association of Patients with SCA1 and other NDDs in the Republic of Sakha (Yakutia). The purpose of this project is to provide practical assistance to patients with neurodegenerative and neuromuscular diseases in traveling on public transport, adaptation in an urban environment and public places, as well as identification by society of special people with neurodegenerative and neuromuscular diseases. Our doctors actively make their personal contribution to the activities of the Association, co-financing from the participants of charity projects makes it possible to purchase medical equipment for examinations and basic necessities that facilitate the stay of patients in the clinic. Patients with SCA1 and other NDDs often express gratitude to the medical staff of the YSC CMP Clinic for their professionalism, careful and sensitive attitude towards patients [2,13].

The principle of respect for personal autonomy. According to Kant's deontological ethics, each person is unconditionally invaluable and he himself is able to determine his own fate: "Each person is an end in itself and, in no case, should be considered as a means to accomplish any task, even if they were tasks for the common good" [21].

Autonomy is deliberate self-government. If we have autonomy, we can make our own decisions based on deliberation. Sometimes we may intend to do something as a result of these decisions, and sometimes we may do these things to implement the decisions. Respect for autonomy is a moral obligation to respect the autonomy of others to the extent that such respect is compatible with equal respect for the autonomy of all potentially affected [1,31,32].

In medicine, respect for autonomy must be strictly observed, and it is closely linked to ethical rules:

- *maintaining confidentiality*. Healthcare professionals promise their patients and clients that they will keep confidential the information they have been entrusted with;
- *informed consent*. Medical professionals promise to consult

with patients and get their consent before providing any medical care;

- *a trusting relationship*. Medical professionals promise a type of communication in which it is possible to find out whether patients want more information or less information that they really want to receive about the prognosis and choice of treatment methods, since some patients prefer to leave decisions to doctors, while others leave the decision to themselves [35,38,44].

We investigated the bioethical aspects of medical and genetic counseling for families burdened with SCA1 in the context of the principle of respect for individual autonomy. The main controversial bioethical issues are:

- what is the priority - personal autonomy or the right of family members to information?
- is the genetic information obtained as a result of DNA testing clinically useful for the patient if the disease is incurable?
- can socio-cultural and ethnopsychological features influence the patient's ability to make informed decisions and truly voluntary decision-making.

Here, a lot depends on the skills of the consulting geneticist, when consulting families with SCA1, the doctor must assess the psychological well-being of the family. Hodgson (2005) argues that genetic counselors are ethically obligated to consider the family as a meaningful unit. This forces them to reconcile the interests of their patients' autonomy with more subtle concerns about the benefits and harm to families [27,34,40]. Moreover, in practice, patients rarely insist on non-disclosure of the results of genetic tests to relatives [30,36]. Rather than using confidentiality towards an individual patient as a default priority, they suggest that genetic information should be conceptualized as family information. According to this so-called “joint accounting” model, disclosure of medically useful genetic information to relatives is the default practice, unless there are compelling reasons to maintain the confidentiality of individual patients [36,41].

Indeed, when consulting patients with SCA1, compliance with the rule of confidentiality within the family seems almost impossible, since family priorities often prevail over personal ones. In addition, the population in small villages is usually aware of SCA1 patients in burdened families, so information about the disease cannot be withheld. As a result, there is some self-isolation of families with an SCA1 positive family member, or a forced change of residence.

We have identified some ethnopsychological aspects that affect the medical and genetic counseling of families with NDDs. To do this, we turned to the characteristics of the Yakuts described earlier by researchers of the mentality of the people.

Bravina R (2008) notes: "... among the Yakuts, "Destiny" fell by lot to each person in accordance with his innate and genealogical parameters. Destiny, as a burden, provided for the whole set of everyday events assigned to a person, successes and failures, personal values. The "top of happiness" is first of all health and longevity, then offspring (especially sons - heirs and successors of the family) ..."[3].

Yakuts and the peoples of the north have developed a special attitude towards diseases, namely the ability not to dramatize certain manifestations of any disease. Yakuts have a sharply negative attitude towards emphasizing or ridiculing physical disabilities or diseases in people. Mercy is a national trait of character [19]. As many researchers have noted, Yakuts showed special, extraordinary pity and care for their sickly or physically handicapped children and nursed them as best they could. They have always been compassionate towards the disabled and the elderly [3, 10, 18,19]. V.L. Seroshevsky wrote: "The crippled, the frail, the sick could count on a certain guardianship. Taking care of them has always been considered the duty of the family. Iitimni (orphan), kumalaan (disabled adults and the elderly) – this is how the Yakuts call those who are dependent on the family – an ancient family institution… According to Yakut concepts, it is a sin to despise iitimni, ... anyone can become one" [18].

Ethnopsychological features, in our opinion, can have a positive impact on the psychological aspects of medical and genetic counseling of hereditary neurodegenerative diseases, including informed consent and making difficult decisions.

The principle of justice. The principle of justice in bioethics seems to be the most controversial and relative. In the field of health ethics, equity obligations are divided into three categories: - equitable allocation of limited resources (distributive justice);

- Respect for human rights (justice based on rights);
- respect for morally acceptable laws (legal justice).

For example, in the context of the allocation of medical resources, there are possible contradictions between several common moral considerations:

- Is it possible to provide sufficient medical care to meet the needs of all who need it, if it is impossible to allocate health resources in proportion to the degree of people's needs for medical care;
- Can health care workers prioritize the needs of so-called "own" patients if they must ensure equal access to health care;
- Should medical professionals provide people with as much choice as possible

to maximize the benefits derived from available resources, to respect the autonomy of the people to whom these resources are provided. All these criteria for the fair distribution of health resources can be morally justified, but not all of them can be fully met simultaneously [31,32].

Let's consider the "principle of justice" on the example of the most common neurodegenerative disease in the Yakut population - spinocerebellar ataxia type 1, which belongs to rare (orphan) hereditary diseases, its prevalence in world populations is 1-4 per 100,000 population. At the same time, Yakutia is a focus of accumulation of SCA1, according to the latest data, 376 patients were registered, the prevalence was 77.6 per 100,000 population [17,33,42].

This hereditary disease is one of the most studied: a dynamic mutation responsible for the manifestation of clinical signs was discovered by molecular genetic methods, the spectrum of phenogenotypic characteristics, population-genetic features were studied in detail, epidemiology and prevalence in Yakutia were studied. It was found that the disease is late-manifesting and is characterized by the phenomenon of anticipation [14,33,42]. The high prevalence of SCA1 in the population undoubtedly causes concern to the health service of Yakutia, measures such as the medical examination of patients, the maintenance of registers for SCA1, the organization of prenatal diagnostics, the creation of an association of patients and SCA1 and other NDDs have been taken. The translation of scientific research into practical medicine, as it turned out, is a very long process, for example, for SCA1 it took more than 20 years [7]. During this time, we investigated the bioethical problems of using gene methods (DNA testing) in practical medicine. When direct DNA diagnosis became a routine clinical analysis, geneticists initially had a paternalistic desire to advise patients prescriptively for mandatory DNA testing of the carrier of the SCA1 mutation. Discussions of bioethical problems of new gene technologies led us to the adoption of ethical rules of medical and genetic counseling, where non-directive, voluntary, informed consent, and alternative informed choice were fixed when making a decision about DNA testing. For the prenatal diagnosis of SCA1, the ethical principle was adopted that prenatal diagnosis is not a method of getting rid of the fetus carrying the SCA1 mutation, but provides a burdened family with a way to choose termination or prolongation of pregnancy through an informed decision. Thus, we strive to comply with the "principle of fairness" of medical care for patients with SCA1.

Conclusion. Applying basic ethical principles as starting points can lead to different answers regarding specific bioethical problems, in our case the moral problems of neurodegenerative diseases. The underlying principles can help doctors and other healthcare professionals make decisions by reflecting on the moral issues that arise at work. As a result of empirical and ethical research, normative solutions are proposed that have far-reaching consequences for society and the future of individuals. Applied bioethical research identifies morally relevant issues, provides facts, describes the actual behavior of stakeholders, and discusses the possibility of intervention to improve the moral quality of clinical practice.

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