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APPROACHES TO PHYSICAL REHABILITATION OF PATIENTS WITH SPINOCEREBELLAR ATAXIA TYPE 1 IN THE CLINIC OF YSC CMP

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The aim of the study was to search for approaches to physical rehabilitation of patients with spinocerebellar ataxia type 1 (SCA1) using kinesitherapy. Gradual activation of motor activity is the main condition for improving the psychological and physical health of patients with SCA1. In our article, physical rehabilitation is specially directed use of physical exercise as a means of treating a disease and improving body functions that are disrupted or lost due to SCA1's neurodegenerative processes.

Keywords: kinesitherapy, physical rehabilitation, spinocerebellar ataxia type 1, Republic of Sakha (Yakutia).

Introduction. Studies of hereditary diseases of the nervous system are becoming more relevant in the world due to severe clinical manifestations, disability and social maladaptation of patients.

In 1991, dynamic mutations, which are the cause of a large group of diseases of the human nervous system, were described, among which polyglutamine diseases with an increase in the number of CAG repeats and various clinical manifestations, such as Kennedy's disease, Huntington's Chorea and spinocerebellar ataxia of types 1, 2, 3, 6, 7 and 17 are distinguished [10].

Spinocerebellar ataxia type 1 (SCA 1) belongs to the group of neurodegenerative diseases with late manifestation. The mutation causing SCA1 is located on the p arm of chromosome 6 and represents a pathological increase in trinucleotide CAG repeats, therefore SCA1 is also classified as a disease with dynamic mutations. The prevalence of SCA1 varies in different populations of the world [5,8].

In the last two decades, scientific and clinical interest in the use of physical exercises for the treatment of mobility problems in persons with neurodegenerative pathology has sharply increased. Advances in basic scientific research suggest neurochemical and neuroplastic changes after physical exertion, an increasing number of high-quality studies document specific aspects of improving mobility after physical exertion [6,7,9,11].

Individuals suffering from type 1 spinocerebellar ataxia are usually demotivated to engage in recreational physical exercise. The most common obstacles are external barriers: the lack of an accessible environment for people with disabilities, feelings of awkwardness and embarrassment in public, anxiety, frustration and anger. Patients with SCA1 are also demotivated by muscle weakness, stiffness, balance disorders, and fear of injury [9].

One of the forms of exercise therapy is kinesitherapy - a new direction in medicine based on the restoration of the human ability to move normally. A person, performing active and passive movements and certain exercises of therapeutic gymnastics, achieves a definitive improvement in his condition. Kinesitherapy as a specialty has a scientific and applied nature, which combines medicine, pedagogy, physiology and biochemistry. It contributes to the psychoemotional and physical comfort of the individual [2].

The aim of the study was to search for approaches to physical rehabilitation of patients with spinocerebellar ataxia type 1 (SCA1) using kinesitherapy.

Materials and methods. The medical examination of patients with SCA1 and the curation of the Republican medical and genetic register of hereditary and congenital pathology has been conducted in the Medical and Genetic Center of the Republican Hospital No. 1 - NMC since 2001, and in 2019, a Center for Neurodegenerative Diseases was organized in the Clinic of the YSC CMP on the order of the Ministry of Health of the Republic No. 01-07/184 dated 02/14/2019. "On the procedure for routing neurological patients suffering from neurodegenerative diseases at the outpatient and hospital stages" [1,3].

The study was conducted within the framework of the research "Exercise therapy in the rehabilitation system of patients with spinocerebellar ataxia type 1 in the Republic of Sakha (Yakutia)", agreed by the local Committee on Biomedical ethics at the YSC CMP (extract from Protocol No. 53 of April 13, 2021)

The study included patients with SCA1 who were treated in the clinic of YSC CMP during 2021. A total of 14 patients were examined, of which 2 were men and 12 were women; 28% of the examined were young people from 20 to 30 years old; 35% were middle-aged (31-50 years); 35% were elderly patients from 51 to 75 years old. All patients had gait disorders to varying degrees. We used the Morse Fall Scale assessment, which is used to assess the probability of falling risk due to the presence of hereditary or acquired risk factors.

Results and discussion. One of the main clinical signs of SCA1 are gait disorders of the patient, due to damage to peripheral nerves, spinal cord roots, loss of sensitivity in the legs. In addition, with cerebellar ataxia, disorders of motor coordination are present, and patients are concerned about loss of balance, frequent falls [5,6]. In our study, gait disorders were assessed: the majority of patients (86%) receiving maintenance

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therapy at the YSC CMP clinic had mild gait disorders. This is due to the fact that patients with severe gait disorders or using a wheelchair rarely come to the YSC CMP clinic due to the lack of conditions (wheelchair lift / elevator).

We analyzed how SCA1 patients move from their self-reports. In our study, only 28% of patients moved by themselves without support, while the majority walked with the help of a cane (35%) and leaning on furniture and walls (35%)

The assessment of falls was carried out using a standard Morse Fall Scale in points, the frequency of falls of patients with SCA1 was estimated for three months. It turned out that half of our patients (50%) suffered involuntary falls in various situations.

Currently, the Russian system of therapeutic/adaptive physical education faces the task of scientifically substantiating the physical rehabilitation of disabled people for their professional and social adaptation. Specialists in the field of rehabilitation of patients with spinal cord disorders distinguish the following forms of physical education:

- independent physical exercises according to methodological instructions, walking, walks in the outside;
- group classes under the guidance of an instructor in medical institutions and rehabilitation centers [4,6]

Another additional method is the use of demonstrational physical exercises in the online Zoom program. So, in 2021, for the first time, we held a webinar for patients with SCA1 on the topic "The importance of physical education for patients with SCA1".

The main conditions for practicing SCA1 physical therapy are: methodical and daily exercise; exercise should bring pleasure to the patient; it is necessary to adhere to the principle "from simple to complex" (Fig.1).

Gradual activation of motor activity is the main condition for improving the psychological and physical condition of patients with SCA1. At the same time, all types of motor activity are effective: physical exercises, walking, dancing, and practicing suitable sports. Significant emotional stresses, physical exercises requiring intense attention and continuous changes in response to changing conditions are contraindicated in patients with SCA1, it is important to avoid overwork and keep your condition under control [9,11].

We have developed a "Self-monitoring Diary" for patients with SCA1 in the form of a notebook, where patients are offered to perform two sets of simple exercises daily, it also contains recommendations for observing the daily schedule, nutrition, graphs for daily control of heart rate and blood pressure (Fig. 2).

Thus, at this initial stage of research, we are searching for approaches and

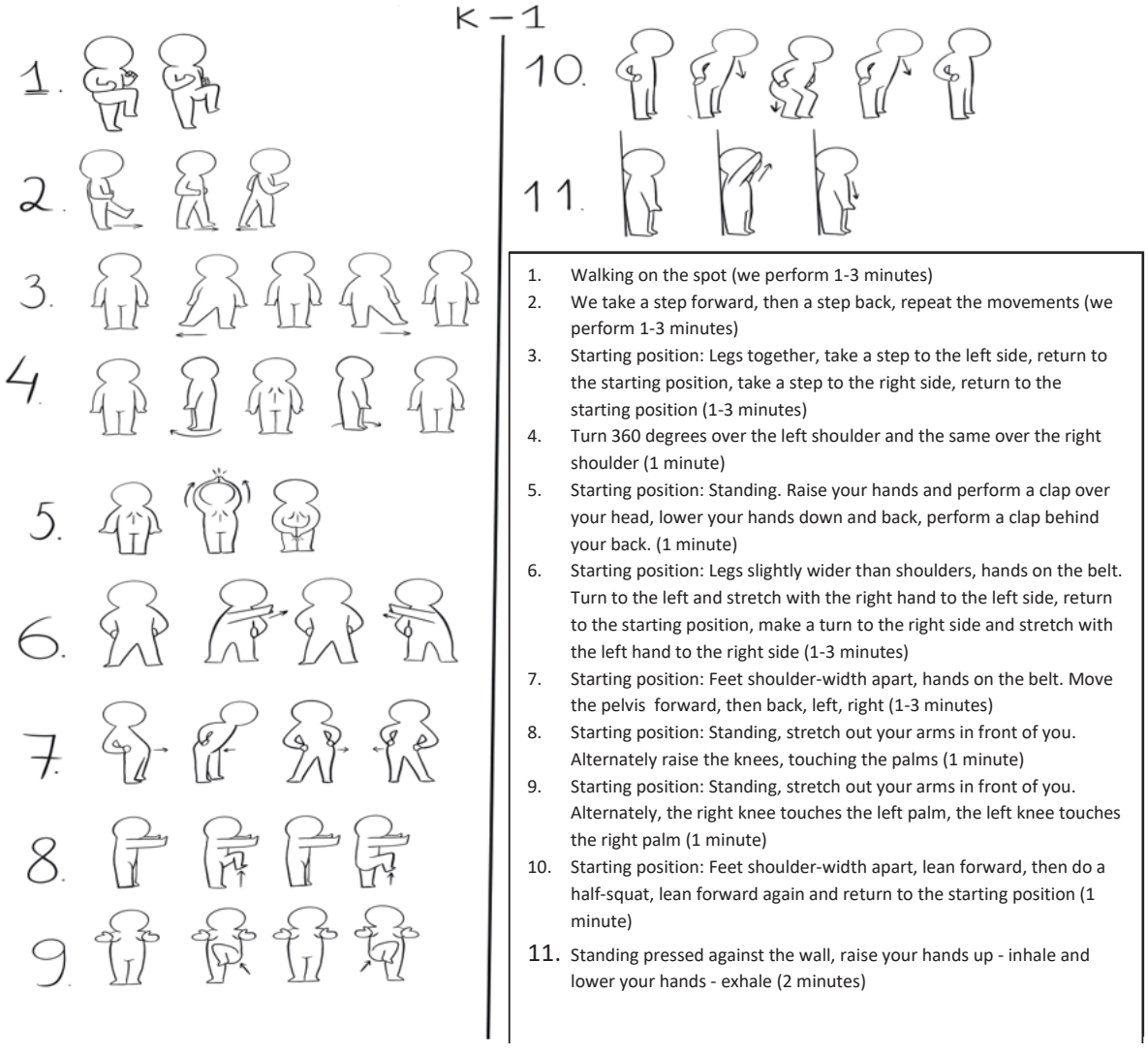

Fig. 1. A set of simple exercises for the development of coordination in patients with SCA1

Fig. 2. Self-monitoring diary for patients with SCA1

methods to the physical rehabilitation of a complex patient group - patients with SCA1. The difficulty lies in the fact that spinocerebellar ataxia type 1 is an incurable and progressive disease of the nervous system. The very awareness of this fact demotivates patients to engage in physical exercise, additional professional psychological assistance is needed for patients receiving maintenance therapy in the clinic.

Conclusion. Taking into account the urgency and sparseness of source material on problems of physical rehabilitation of patients with persistent and progressive health disorders, in the article we describe the first results of kinesitherapy treatment on patients with SCA1. In our article, physical rehabilitation is specially directed use of physical exercise as a means of treating a disease and improving body functions that are disrupted or lost due to SCA1's neurodegenerative processes. At the first stages of our research, we set the tasks of using simple sets of physical exercises so that, firstly, they have a health-improving and restorative effect on the body; secondly, they strengthen weakened muscles, improve coordination of movements. At the next stage, patients with SCA1 will be divided between case and control groups, we will also apply research methods to develop scientific justifications for physical rehabilitation: pedagogical experiment, questionnaire, stabilometry, goniometry, dynamometry, etc.

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