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FEATURES OF THE COGNITIVE SPHERE IN PATIENTS WITH PROGRESSIVE MYODYSTROPHIES

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Summary: Progressive myodystrophies are a generalized name for hereditary diseases of the muscular system, caused by impaired contractility of muscle fibers and manifested by muscle weakness, a decrease in the range of active movements, decreased tone and atrophy. The medical and psychological significance of these diseases is due to the severity of the course and the high level of disability with a significant limitation on the life expectancy of patients.

Key words: progressive muscular dystrophy, cognitive activity, muscle weakness, dystrophin gene, psychological significance.

Резюме: Прогрессирующие миодистрофии — обобщенное название наследственных заболеваний мышечной системы, обусловленных нарушением сократимости мышечных волокон и проявляющихся мышечной слабостью, уменьшением объема активных движений, снижением тонуса и атрофией. Медико-психологическая значимость этих заболеваний обусловлена тяжестью течения и высоким уровнем инвалидизации со значительным ограничением продолжительности жизни больных.

Ключевые слова: прогрессирующая мышечная дистрофия, когнитивная деятельность, мышечная слабость, ген дистрофина, психологическое значение.

Currently, the number of patients with dysfunctions of the musculoskeletal system has increased significantly. Among them, children and adolescents with hereditary myopathies make up a significant proportion. The psychology of children with musculoskeletal disorders is a branch of correctional psychology. Numerous works by clinicians, teachers, and psychologists are devoted to the problems of studying, training, education, treatment and comprehensive rehabilitation of these patients [4,5]. Despite the fact that extensive empirical material has been accumulated on the study of the mental development of children with cerebral palsy and other disorders of the musculoskeletal system, the problems of development of children with hereditary myopathies are not sufficiently covered.

Hereditary myopathies are a generalized name for hereditary diseases of the muscular system, caused by impaired contractility of muscle fibers and manifested by muscle weakness, a decrease in the range of active movements, decreased tone and atrophy [6]. The medical and psychological significance of these diseases is due to the severity of the course and the high level of disability with a significant limitation on the life expectancy of patients.

This group of diseases is usually distinguished: congenital structural myopathies, progressive muscular dystrophies and metabolic (mitochondrial) myopathies.

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Various forms of hereditary progressive disorders of the musculoskeletal system, described in all countries of the world, occur with a frequency of 1: 25,000 population. The frequency of congenital structural myopathies is 15.3% of all cases of neuromuscular diseases in children in Russia and neighboring countries. The incidence of congenital muscular dystrophies was 4.65 per 100,000 people in Italy.

Diagnosis of myopathies, like other movement disorders, is quite complex, in many cases a muscle biopsy is necessary, and a family history is almost always required.

Currently, the problem of hereditary movement disorders (including myopathies) is considered mainly from a clinical aspect, therefore the issues of psychological and social adaptation of this category of patients are very significant.

In this regard, fundamental changes are needed in the organization of psychological, diagnostic and rehabilitation activities in this area, without which medical and psychological care will not be ready for the challenges of our time.

Of particular importance is the study of the characteristics of cognitive activity of children and adolescents with congenital myopathies, the selection of methods for differentiated neuropsychological diagnostics and the development of programs for adequate psychological and pedagogical correction to improve the quality of life of these patients.

In the works of domestic and foreign psychologists, there is practically no assessment of the formation and development of cognitive function in children and adolescents with hereditary myopathies.

In modern domestic and foreign literature on psychology, the neuropsychological aspect of the course of the disease has not been studied. Also, the issues of psychological and pedagogical correction and social adaptation of children and adolescents with this disease have not been studied. There are no studies describing the relationship between the characteristics of the mental activity of children and clinical indicators in various forms of hereditary myopathies, and knowledge of the developmental characteristics of children with varying degrees of severity of the disease makes it possible to organize effective differentiated care for them.

In a study by Bresolin et al., it was found that 32% of patients with Duchenne disease have a Wechsler coefficient less than 75 and only 24% have an IQ corresponding to the population level. Pleiotropy (the ability of one gene to influence multiple phenotypic traits) of affected genes in muscular dystrophies has been suggested. A. Emery et al. studied heterogeneity in Duchenne muscular dystrophy, comparing a group of boys with severe mental retardation and a group with no severe cognitive impairment.

In patients with severe mental retardation, a later onset of the disease was noted, and they later developed the need to use a wheelchair; they had a less pronounced decline in creatine phosphokinase levels with age. Thus, the severity of muscle damage did not correspond to the level of intellectual impairment. Other studies did not establish a relationship between the severity of mental retardation and the size of the deletion in the dystrophin gene. Recently, the role of dystrophin-glycoprotein complex disorders in the development of a

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spectrum of brain abnormalities from mild cognitive impairment, not manifested by visible brain defects, to severe neuronal migration disorders has been recognized [8].

In general, progressive muscular dystrophies (with the exception of progressive Duchenne muscular dystrophy) are not accompanied by a decrease in intelligence. Patients are critical of their condition. The literature describes pronounced emotional disturbances in these patients, namely increased irritability and depressed mood. Basically, children and adolescents with progressive muscular dystrophies are educated according to the mass school program.

The study of mental changes in patients with hereditary myopathies was a new stage in the study of the disease. The first domestic scientists to work in this direction were L. O. Badalyan, P. A. Temin, Yu. E. Veltishchev [6].

Opinions about the presence of specific disorders of higher mental functions in hereditary myopathies are contradictory, however, some authors point to changes in the psyche, the severity of which varies greatly in different nosological units of hereditary myopathies. Thus, unlike patients with congenital structural myopathies, children with muscular dystrophy often lag behind in psycho-speech development.

Currently, the study of disorders of higher mental functions in children and adolescents with hereditary myopathies is actively continuing.

In the studies of Ippolitova M.V., Kalizhnyuk E.S., Simonova N.V. Mamaichuk I.I., Levchenko I.Yu. notes that along with violations of fine and gross motor skills, difficulties in mathematics and writing, children with motor pathology have a clear functional deficiency of the spatial factor.

The works of I. I. Mamaychuk provide a comprehensive analysis of the characteristics of the cognitive activity of children with cerebral palsy and other forms of motor impairment. Underdevelopment of spatial perception is observed in patients with various forms of musculoskeletal disorders [5].

Together with the pathology of the motor-kinesthetic functional system, sensory, mental and speech disorders observed in this category of patients play a significant role in impaired perception [3, 5].

In addition, the studies of I. I. Mamaichuk revealed that the severity of the motor defect is not the determining factor in the impairment of sensory-perceptual activity in patients with pathology of the musculoskeletal system.

Research by psychologists and teachers emphasizes the presence of difficulties in performing counting operations, as well as underdevelopment of visual-spatial functions, decreased attention span and memory in children with cerebral palsy and other diseases of the musculoskeletal system [3,4].

This poses the task of studying the state of higher mental functions in patients with hereditary motor disorders using neuropsychological examination methods, which will allow

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us to compare the detected cognitive deficiency with the nosological form, the age of the patients and other factors that can also influence changes in the psyche.

The organization of spatial representations, as is known, is provided by parts of the brain located in the tertiary zones of the cortex, located in the posterior parts of the brain on the border between the occipital, temporal and postcentral regions of the hemisphere. They constitute the overlap area of the cortical sections of the visual, auditory, vestibular and skin-kinesthetic analyzers. The centers of these zones are the 39th and 40th Brodmann areas, or the inferior parietal region. In addition, the zone that provides spatial synthesis includes the temporo-occipital formations of the 37th and 21st fields. All these fields perform mainly associative functions. The fibers coming to them come from the associative nuclei of the thalamus and carry information already generalized at lower levels. These zones are formed only in humans and mature later than other zones of the posterior cortex, and begin to fully function only by the age of seven [2].

References

1. Лурия А.Р. Высшие корковые функции человека и их нарушения при локальных поражениях мозга/ А.Р. Лурия. - СПб.: «Питер», 2008.
2. Лурия А.Р. Основы нейропсихологии: учебное пособие/ А.Р. Лурия. -М.: Академия, 2006. - 384 с.
3. Мамайчук И.И. Нейропсихологическое исследование гностических процессов у детей с различными формами ДЦП / И. И. Мамайчук // Невропатология и психиатрия. - 1992. - №4. - С.42-44.
4. Мамайчук И.И. Психологическая помощь детям с проблемами в развитии / И.И. Мамайчук - СПб,: Речь, 2001.- 220 с.
5. Мамайчук И.И. Психология дизонтогенеза и основы психокоррекции (на материале изучения детей и подростков с двигательными нарушениями): автореф. дис. / Мамайчук Ираида Ивановна. - СПб., 2004. -39 с.
6. Наследственные болезни нервной системы: руководство для врачей; под ред. Ю.Е Вельтищева, П.А.Темина. - М.: Медицина, 1998, С. 200-235.
7. Kamoshita S. Congenital muscular dystrophy as a disease of the central nervous system / S.Kamoshita, Y.Konishi, M.Segawa, et al // Arch Neurol. - 1976. -V.33. - P.513-516.
8. Waite A. The dystrophin-glycoprotein complex in brain development and disease / A.Waite, S.C.Brown, D.J. Blake // Trends Neurosci. - 2012. - V.35 (8). -P.487-496.