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Electroneuromyography in pediatric practice (literature review)

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ABSTRACT

One of the most informative diagnostic methods for diseases of the spinal cord and peripheral nervous system is electromyography, a diagnostic technique based on the registration and evaluation of electrical potentials that occur during muscle contraction or peripheral nerve activation in response to electrical stimulation of the nerves. This method is characterized by high variability in the obtained potential parameters: amplitude, latency, morphology, the ratio of these parameters between responses recorded at different stimulation points, as well as conduction velocities along motor and sensory fibers of peripheral nerves. The performance of electromyography in children is associated with several additional complicating factors, such as the high sensitivity of children to the procedures, the difficulty in ensuring patient immobility, and the dependence of the obtained data on the age of the subject. This study involved a review of recent domestic and international literature on the use of electromyography in pediatric practice. Currently, two main electrophysiological methods are used: stimulation electromyography and needle electromyography. In the first case, the conduction of signals along peripheral nerves is assessed, while in the second, the condition of individual muscle fibers is evaluated. During the examination, diseases of the muscles, peripheral nervous system, and spinal cord can be identified; it is also possible to compare the obtained results to assess inter-lateral asymmetry and track the dynamics of pathological processes over time. Electromyography is a highly informative diagnostic tool for neuromuscular system diseases in children; however, when conducting the study, it is important to take into account the age-related features of nervous system development and adapt the procedure according to the physiological and behavioral characteristics of the subjects.

Keywords: pediatrics, electroneuromyography, needle electromyography, diabetic polyneuropathy, muscular hypotonia syndrome

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Электронейромиография в педиатрической практике (обзор литературы)

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РЕЗЮМЕ

Актуальность изучения возможностей применения электронейромиографии в педиатрической практике обусловлена широким спектром нервно-мышечных заболеваний, при которых данная методика является высокоспецифичной и информативной. Электронейромиография — это один из наиболее информативных методов диагностики заболеваний спинного мозга и периферической системы. Метод основан на регистрации и оценке электрических потенциалов, возникающих при сокращении мышцы или активации периферического нерва в ответ на стимуляцию нервов электрическими импульсами. Электронейромиография отличается высокой вариабельностью показателей полученных потенциалов: их амплитуды, латентности, морфологии, соотношения данных показателей между ответами, полученными в различных точках стимуляции, а также скоростей проведения по моторным и сенсорным волокнам периферических нервов. Проведение электромиографии у детей сопряжено с рядом дополнительных осложняющих факторов, таких как высокая чувствительность детей к производимым манипуляциям, сложность обеспечения неподвижности пациента и зависимость полученных данных от возраста обследуемого. В ходе данного исследования был проведен анализ отечественной и зарубежной литературы последних лет на тему возможности использования электромиографии в педиатрической практике. В настоящее время используются два основных электрофизиологических метода исследования: стимуляционная электронейромиография и игольчатая миография. В первом случае оцениваются показатели проведения по периферическому нерву, во втором — состояние отдельных мышечных волокон. При проведении исследования могут быть выявлены заболевания мышц, периферической нервной системы, спинного мозга, миогенные расстройства; возможно сравнение получаемых показателей для оценки межсторонней асимметрии, динамики патологического процесса во времени. Электромиография является высокоинформативным методом диагностики заболеваний нейромышечной системы у детей, тем не менее при проведении исследований нужно учитывать возрастные особенности развития нервной системы, адаптировать проведение методики с учетом физиологических и поведенческих характеристик обследуемых.

Ключевые слова: педиатрия, электронейромиография, игольчатая электромиография, диабетическая полинейропатия, синдром мышечной гипотонии

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BACKGROUND

Electrophysiological tests, such as evoked electroneuromyography (ENMG) and needle electromyography (EMG), are used to diagnose a wide range of disorders of the muscles, peripheral nervous system, and spinal cord, including in pediatric practice. These methods offer a number of advantages, including the ability to obtain quantitative data that allow for an objective assessment of the neuromuscular system, comparison of results with age-specific norms, and monitoring changes in parameters during follow-up examinations.

In our work at the Clinical Hospital of St. Petersburg State Pediatric Medical University, we evaluated the potential for using electrophysiological examination methods in pediatric practice. An analysis of recent domestic and international literature describes the possibilities and limitations of using EMG in pediatric neurology for patients of various age groups [5, 6].

During the studies, a number of parameters can be obtained: for example, during ENMG, one can record the amplitude, duration, latency, and shape of motor and sensory responses; calculate conduction velocities along peripheral nerves; and assess the stability of late phenomena; during needle EMG, pathological spontaneous activity of muscle fibers can be identified, and the amplitude and duration of motor unit potentials, the number of turns and phases in their structure can be assessed, as well as an analysis of the interference pattern and turn-amplitude analysis can be performed at maximum contraction of the muscle under examination [3]. ENMG can be used to diagnose conditions such as mono- and polyneuropathies, myasthenia gravis, tunnel syndromes, plexopathies, neural amyotrophies, and radicular syndromes; needle EMG — for diagnosing lesions of primary muscular and denervation origin [9, 19, 27].

Despite the high diagnostic value and informative nature of the EMG method, its application in pediatric practice is associated with certain difficulties due to the physiological, anatomical, and psychological characteristics of childhood, including variability in normal values, difficulties in ensuring the child's immobility during the examination, as well as children's increased sensitivity to external manipulation.

Physiological characteristics include incomplete myelination of peripheral nerves, active synaptogenesis, and the growth, maturation, and branching of axons. The presence of a myelin sheath covering the axons of nerve fibers is necessary to achieve sufficient nerve conduction velocity. Thus, incomplete myelination accounts for the characteristics of peripheral nerve conduction velocity and the increase in response latency. As a rule, a rapid increase in conduction velocities is observed after birth; velocity parameters reach values similar to those recorded in adult patients by approximately 3–5 years of age, while certain changes, such as bimodality (the presence of two negative peaks) in sensory responses, continue to be observed until 15–16 years of age [16, 33]. When performing needle EMG, it should be noted that in cases of primary muscle lesions, instead of the interference pattern filled with low-amplitude potentials typical in adult practice, motor unit potentials with normal or increased amplitude and reduced duration may be recorded. These changes may be associated with hypertrophied muscle fibers. Broad, high-amplitude, polyphasic potentials similar to those observed in denervation lesions may also be recorded; in such cases, attention should be paid to the presence of both broad and narrow polyphasic potentials — this feature is more characteristic of the primary muscle pattern [37].

Another important factor to consider when conducting the study is the anatomical characteristics of the patients. In studies of the adult population, the distances between the stimulation site and the recording site are fixed and standardized in accordance with international recommendations, ensuring high reproducibility of results [17, 24, 26]. In children, especially younger ones, these distances often vary due to anatomical characteristics: smaller body size, lower muscle mass, and body proportions that differ from those of adult patients. This requires a flexible approach to selecting stimulation and recording sites, as well as adjusting measurements based on age and the size of the area being examined. The lack of fixed standards for children creates additional complexity in interpreting results and requires the use of age-specific reference data. Additionally, interelectrode distances may be reduced, as may the distance between the stimulating and recording electrodes. Anatomical landmarks may be used during the study [37, 40].

It should be noted that the examination may cause pain and discomfort in the area of stimulation or needle electrode insertion. Unlike adult patients, most of whom tolerate EMG satisfactorily, unpleasant sensations may cause anxiety and motor activity in the subjects, leading to an increase in the number of recording artifacts, as well as the inability to perform procedures that require patient cooperation; furthermore, communication with young children may be impossible. When conducting the examination, it is recommended to establish rapport with the child: for example, it may be helpful to engage the child in the procedure by explaining that the sounds recorded during EMG are "muscle sounds" captured by a microphone. It is recommended to avoid terms like "needle" or "needle electrode" as they carry negative

connotations. Phrases such as “recording electrode”, “microphone”, “antenna”, or others that will be perceived more positively by the child should be used. A higher quality of the examination can be achieved if a staff member or parent present distracts the child with videos, games, or books [31]. When examining patients of preschool and younger ages, parents may be invited to be present during the procedure and hold the child in their arms; however, it is necessary to explain that the child may become anxious and cry during the examination [37]. The possibility of sedating patients should be considered, as a number of publications indicate an improvement in data quality when sedation is used [18, 25], while others show that electromyography can be performed with sufficient accuracy without sedation [22].

Thus, the recording of electrophysiological parameters presents certain challenges, and a correct interpretation of the data is essential for reaching a reliable conclusion. A number of scientific studies describe conduction parameters recorded in a population of healthy children [12, 18, 20, 38, 40], noting the high variability of the data — each individual electrophysiological laboratory is recommended to develop its own reference tables or, at a minimum, cite the source according to which the recorded values are interpreted [14, 40, 44].

The use of evoked EMG and needle EMG techniques is quite common in clinical practice. The most common form of polyneuropathy in childhood is diabetic polyneuropathy. A significant number of articles describe changes in electrophysiological parameters in children with diabetes mellitus — a disease whose prevalence is steadily increasing [8, 29]. During evoked potential studies in this population, changes such as reduced motor conduction velocities [2, 23, 30] and reduced amplitudes of motor and sensory responses [2, 7] were observed. There is a challenge in determining the scope of the examination sufficient to draw a reliable conclusion — the need to examine both motor and sensory fibers of at least one lower limb has been described, while the required number of nerves examined and the necessity of assessing nerve conduction parameters in the second lower limb and upper limbs vary [10, 15]. The literature describes a correlation between increased variability in blood glucose levels and a decrease in nerve conduction velocity [32, 41]. It has also been noted that, regardless of the duration of the disease, significantly pathological changes on ENMG are observed much less frequently than during confocal microscopy with assessment of corneal nerve fiber function. Conduction parameters in the population of children with diabetes mellitus are predominantly within the normal range but are reduced relative to those recorded in the population of healthy children [34].

The use of ENMG is not limited to the examination of children with diabetes mellitus. The examination of patients with hereditary neuromuscular diseases, metabolic disorders, rheumatic diseases, and neuropathies includes ENMG. A number of publications from 2020 and later are devoted to myographic parameters in children who have had COVID-19 followed by the development of Guillain–Barré syndrome. In a cross-sectional study involving ENMG in 125 patients reporting complaints of respiratory failure, paralysis of the upper and/or lower extremities, and gait disturbances, it was found that Guillain–Barré syndrome is characterized by an electrophysiological pattern of axonal damage to peripheral nerves, with a predominance of reduced amplitude in motor and sensory responses. Furthermore, the axonal type of lesion was associated with a less favorable prognosis for recovery than the demyelinating type [21]. Similar findings were reported in descriptions of individual clinical cases: changes of both the axonal [13] and demyelinating types [35] were noted. In a number of cases, a multisystem inflammatory syndrome associated with COVID-19 was identified in children, in which, in addition to peripheral nervous system involvement, central nervous system involvement was noted, and psychoses and epileptic seizures were described [39].

One of the most common reasons for performing an electrophysiological examination on an infant under one year of age is neonatal brachial plexus injury, which results from overstretching of the brachial plexus nerve fibers during childbirth. This condition may manifest as either a temporary decrease in arm motor function or the development of permanent complete paralysis of the upper limb. Typically, the C_5 and C_6 roots are affected (in about 80% of cases), but in some patients the C_7 root may also be involved, and rarely the C_5–T_1 roots (pamplexopathy) [28]. Depending on the extent of brachial plexus involvement, changes may vary — from minimal deviations in conduction parameters to a complete absence of responses. A combined study using both evoked EMG and needle EMG is recommended; however, it should be noted that performing needle myography is complicated by the inability to establish productive contact with the patient. Furthermore, the predominance of unilateral lesions allows for a comparative analysis of electrophysiological parameters between the affected and healthy limbs, which, given the high variability of conduction parameters in young children, significantly improves the quality of the study [42, 43].

In the practice of a pediatric electrophysiologist, it is common to encounter cases where a young child with muscular hypotonia syndrome is referred for examination.

It should be noted that the causes of this condition can vary. Given the discomfort associated with the procedure, which causes anxiety in the patient and reduces the quality of the data obtained, it is recommended to first employ other diagnostic methods (medical history review, physical examination, imaging studies, genetic/laboratory tests) followed by the development of a strategy for performing myography [1, 4, 11, 36].

CONCLUSION

The use of EMG in pediatric practice is becoming increasingly common: the quantitative data obtained are objective and can be used to compare readings between sides, assess changes over time, and monitor the effectiveness of treatment. This method can be used in children of any age. The examination allows for the differential diagnosis of a wide range of muscular and neurological disorders.

ADDITIONAL INFORMATION

Author contribution. All authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study.

Competing interests. The authors declare that they have no competing interests.

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