ELECTRONEUROMYOGRAPHIC (ENMG) CHANGES IN CONGENITAL MYOPATHIES AND THEIR ROLE IN DIAGNOSIS

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Abstract: This study aims to assess the diagnostic value of electroneuromyographic (ENMG) changes in congenital myopathies. ENMG studies allow us to study the functional state of muscles and peripheral nerves by determining their electrical activity. The results of the study showed that congenital myopathies are characterized by low amplitude of muscle potentials, short-duration motor unit potentials, and normal nerve conduction. These features allow using ENMG as an effective method for early detection of congenital myopathies and their differentiation from neuropathies.

Keywords: Congenital myopathy; ENMG; Muscle activity; Nerve conduction.

Objective: To identify electroneuromyographic (ENMG) changes in patients with congenital myopathies, assess their dependence on clinical symptoms, and determine the value of this method in early diagnosis of the disease based on ENMG indicators.

Research object and methods:

The object of the study was 20 patients of different ages with suspected congenital myopathy and a control group of 10 healthy individuals. All participants underwent a clinical and neurological examination.

The research methods included electroneuromyography (ENMG). The amplitude, duration, polyphasicity and nerve conduction velocity of muscle potentials were assessed using ENMG. The obtained data were compared with the indicators of a healthy control group and statistically analyzed.

Discussion: The conducted electroneuromyographic (ENMG) studies showed a significant decrease in the electrical activity of muscle fibers in patients with congenital myopathy. Mainly low-amplitude, short-duration motor unit potentials and normal nerve conduction were observed. These changes are associated with primary damage to muscle tissue, and the absence of signs of neuropathy is an important criterion confirming the diagnosis of congenital myopathy.

Thus, the results of ENMG are important as a reliable instrumental method for early detection of muscle damage in congenital myopathies, differentiation of the disease from peripheral nerve diseases, and clarification of the diagnosis.

Results:

The study involved 20 patients (mean age - 8) with suspected congenital myopathy and 10 healthy children. According to the results of ENMG, the amplitude of the motor unit potential in patients with

congenital myopathy was 0.3 ± 0.1 mV, which was significantly lower than the indicator in the control group (1.2 ± 0.3 mV) ($p < 0.01$).

The duration of the potentials was 4.8 ± 1.1 ms, while in healthy subjects this indicator was 9.2 ± 1.5 ms ($p < 0.05$). The proportion of polyphasic waves was on average 18% in patients and 42% in the control group.

Nerve conduction velocity (motor nerves) was 52 ± 4 m/s in patients and 54 ± 3 m/s in healthy controls, with no significant difference ($p > 0.05$).

These results indicate that muscle dysfunction is associated with a primary myogenic process, while nerve conduction is preserved.

Recommendations:

1. It is recommended that all patients suspected of congenital myopathy undergo ENMG examination at the initial examination stage.

2. Evaluation of ENMG results in conjunction with clinical signs increases diagnostic accuracy.

3. ENMG monitoring can be used to assess the effectiveness of rehabilitation and treatment processes.

Conclusion: Electroneuromyographic (ENMG) studies in patients with congenital myopathy showed a significant decrease in the electrical activity of muscle fibers. A decrease in the amplitude of motor unit potentials, a decrease in their duration, and a decrease in the proportion of polyphasic waves are changes characteristic of primary damage to muscle tissue. The preservation of nerve conduction is explained by the myogenic, rather than neuropathic, nature of the disease. The ENMG method is a reliable and informative instrumental method for early diagnosis of congenital myopathies.

References

1. Shanina, E. (2022). *Electrodiagnostic evaluation of myopathy*. StatPearls [Internet]. (Updated review on needle EMG and interpretation in myopathies).
2. Wijntjes, J., et al. (2024). *Comparison of muscle ultrasound and needle EMG in patients suspected of a neuromuscular disorder*. Muscle & Nerve, 2024. (Concurrent ultrasound vs needle-EMG study).
3. Zhang, H., et al. (2024). *Congenital myopathies: pathophysiological mechanisms and diagnostic approaches*. Journal of Translational Medicine, 2024. (Review covering genetics, pathology, and diagnostic workup including EMG).
4. Zhang, Y., et al. (2022). *Clinical and genetic features of infancy-onset congenital myopathies: implications for diagnosis (incl. EMG)*. BMC Pediatrics, 2022.
5. Dowling, J. J., & Nishino, I. (2020). *Muscle biopsy, genetics, and biomarkers in congenital myopathies: an integrated diagnostic model*. Seminars in Pediatric Neurology, 35, 100847. (Contextualizes EMG within multimodal diagnosis).