



Current Views on the Diagnosis and Treatment of Juvenile Myasthenia Gravis

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Abstract: Myasthenia gravis is an autoimmune neuromuscular disease. In people with myasthenia gravis, the nerves and muscles cannot communicate normally. This is caused by abnormal antibodies that disrupt the communication between muscles and nerves. This disorder causes varying degrees of muscle weakness. The name of the disease comes from the Greek words myo or "muscle", asthenos or "without strength" and gravis, meaning "heavy".

Key words: juvenile myasthenia gravis, autoimmune neuromuscular disease.

Introduction. Myasthenia gravis is rare: about 10 out of a million people are diagnosed each year, and only 10 percent of them are children. The disease occurs in all age groups, ethnicities, and both sexes. When the condition is diagnosed in a child, the most common form is called juvenile myasthenia gravis (JMG). The juvenile form is more variable in its manifestations and can mimic other diseases, making it difficult to diagnose. For example, autoantibodies in the blood are more difficult to detect in children than in adults.



Fig.1 Juvenile myasthenia gravis

In other patients, more muscles are affected, and additional symptoms may include double vision, clumsiness, falling over, difficulty speaking or swallowing, shortness of breath, and rapid fatigue during play. Sometimes children are mistakenly thought to be lazy, uncoordinated, or even unmotivated.

Causes of myasthenia gravis in children

The cause of the development of UMH in children is unknown. In people with UMH, nerves and muscles cannot communicate normally. This communication process usually occurs with a molecule (acetylcholine) that travels from the nerves to the muscle cells. In UMG, abnormal antibodies attach themselves to the muscle and prevent acetylcholine from working properly. Eventually, the lack of nerve transmission leads to muscle weakness. These abnormal antibodies are produced by the immune system. The immune system, which normally protects the body from foreign organisms, does not work properly in patients with SMH. Although scientists do not yet fully understand why, it is thought that the thymus gland, an organ located in the chest, may not properly affect the immune system. Many patients with UMH demonstrate thymic dysplasia or abnormal growth of the thymus. Clinical improvement can also be demonstrated with removal of the thymus gland in most patients who undergo surgery.

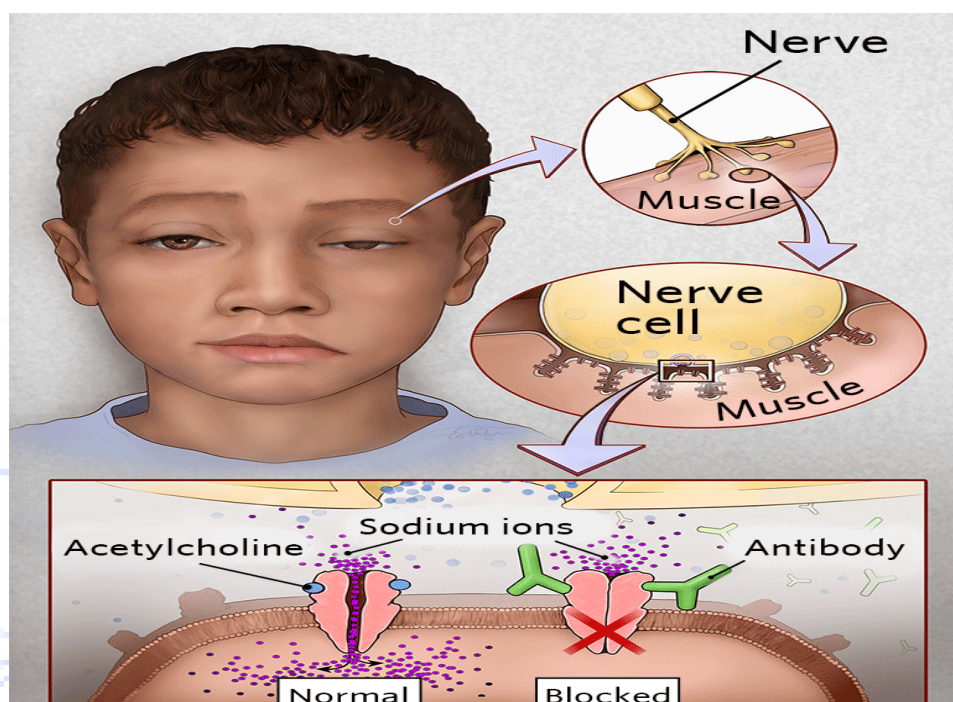


Fig.2 Diagram showing the connection between nerves and muscles in myasthenia gravis

This diagram demonstrates the connection between nerves and muscles. The clinical manifestations of myasthenia gravis, including symptoms such as ptosis, arise from poor communication between nerves and muscles at the cellular level. This diagram shows the detail of the "nerve-muscle connection," which is the space between nerves and muscles. Normally, a molecule called acetylcholine (blue dots) is released from the nerve cell and attaches to a receptor on the muscle cell (shown in pink). This allows sodium (purple) to enter the muscle and initiate muscle contraction. In myasthenia gravis, abnormal antibodies (green) interfere with acetylcholine binding, stopping the normal process of muscle-nerve communication.

Myasthenia gravis testing and diagnosis

JMG is diagnosed through a comprehensive physical examination and several diagnostic tests. Each case is different, so the tests used may vary.

At CHOP, your child will have a blood test to check for irregular antibodies. If this test is positive, the diagnosis of JMG will be confirmed. If the test is negative, other tests may be done, including:

Diagnosis

- Nerve stimulation testing: the connection between nerves and muscles is tested by repeated nerve stimulation.
- Electromyography (EMG): single fiber electromyography measures the response of an individual muscle fiber to electrical impulses.
- Intravenous tensilon testing (edrophonium): intravenously administered edrophonium chloride can improve myasthenia gravis symptoms for a short period of time. This test is positive if there is a significant improvement in symptoms after administration. This should be done in a carefully controlled setting. A CT or MRI scan may also be done to obtain images of the chest. These images can help plan your child's treatment and rule out the presence of a very rare tumor called a thymoma.

Treating myasthenia gravis in children

Non-surgical treatment. There are several medications that can help manage the symptoms of UMH. The two main types of medications are:

- Acetylcholinesterase inhibitors are the first-line treatment. They increase the availability of a molecule (acetylcholine) that helps nerves and muscles communicate. This medication is only marginally effective for severe symptoms. The most commonly prescribed medication is called pyridostigmine, also known as Mestinon. Side effects include diarrhea and muscle cramps.
- Immunosuppressive therapy (oral steroids and/or intravenous immunoglobulin) is used both to relieve symptoms and to stabilize patients before surgery. Physicians try to limit the amount of time patients are exposed to these drugs in order to minimize side effects in children over time. For example, steroids can lead to stunted growth and obesity. In low doses, steroids are better tolerated. If higher doses are needed to treat symptoms, other immunomodulatory drugs may be considered.

Surgical treatment

Although medications can be very effective in treating myasthenia gravis, we believe that a combination of medication and surgery offers the best chance of remission. Surgical treatment includes removal of the thymus gland, a procedure known as a thymectomy.

The goal of surgery is to achieve remission, improve symptoms, and reduce medication use. However, its effect can sometimes be seen for months to years after surgery.

Conclusions: Thus, with treatment, muscle weakness often gets much better. Medications can help improve the connection between nerves and muscles and make muscles stronger. Other medications prevent your body from producing as many abnormal antibodies. These medications can have serious side effects, so they should be used with caution. There are also treatments that filter out the abnormal antibodies from the blood or add healthy antibodies from donor blood. Sometimes surgery to remove the thymus gland helps.

Literature:

1. Sadriddin Sayfullaevich Pulatov. (2022). Verticalization of Patients with Severe Acquired Brain Injury: A Randomized Pilot Study. Eurasian Medical Research Periodical, 9, 46–48.
2. Khamdamov B.Z. Indicators of immunocytocine status in purulent-necrotic lesions of the lower extremities in patients with diabetes mellitus.//American Journal of Medicine and Medical Sciences, 2020 10(7) 473-478 DOI: 10.5923/j.ajmm.2020.- 1007.08 10.

3. M. I. Kamalova, N.K.Khaidarov, Sh.E.Islamov, Pathomorphological Features of hemorrhagic brain strokes, Journal of Biomedicine and Practice 2020, Special issue, pp. 101-105
4. Kamalova Malika Ilkhomovna, Islamov Shavkat Eriyigitovich, Khaidarov Nodir Kadyrovich. Morphological Features Of Microvascular Tissue Of The Brain At Hemorrhagic Stroke. The American Journal of Medical Sciences and Pharmaceutical Research, 2020. 2(10), 53-59
5. Khodjieva D. T., Khaydarova D. K., Khaydarov N. K. Complex evaluation of clinical and instrumental data for justification of optive treatment activites in patients with resistant forms of epilepsy. American Journal of Research. USA. № 11-12, 2018. C.186-193.
6. Khodjieva D. T., Khaydarova D. K. Clinical and neuroph clinical and neurophysiological ch ogical characteristics of teristics of post-insular cognitive disorders and issues of therapy optimization. Central Asian Journal of Pediatrics. Dec.2019. P 82-86
7. Sadriddin Sayfullaevich Pulatov. (2022). Efficacy of ipidacrine in the recovery period of ischaemic stroke. World Bulletin of Public Health, 7, 28-32.
8. Sadriddin Sayfullaevich Pulatov. A Randomised Pilot Study Of Cardiovascular Patients With Hemispheric Ischaemic Stroke Against A Background Of Early Verticalisation- Web Of Scientist: International Scientific Research ..., 2022

