Ocular Myasthenia Gravis

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A Patient’s Guide

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Diagnosis</th>
<th>Treatment</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Nerve</th>
<th>MUSCLE CELL</th>
<th>Neuromuscular Junction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal Nerve Transmission</td>
<td>Acetylcholine</td>
<td>Acetylcholine Receptor</td>
</tr>
<tr>
<td>NERVE ENDING</td>
<td>Antibody Against Receptor</td>
<td></td>
</tr>
<tr>
<td>Myasthenia Gravis</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Acetylcholine

MUSCLE CELL
What is myasthenia gravis and what causes it?

Myasthenia gravis is a condition that causes weakness of specific muscles in the body. Normally nerves send a signal to muscles using a chemical called acetylcholine, which tells the muscles when to move. In patients with myasthenia gravis, the body’s immune system mistakenly interferes with the muscles’ receptors for acetylcholine. When these receptors cannot work properly, the affected muscles tire easily. The amount of weakness typically fluctuates and may be worse at the end of the day.

Nerves release a chemical called acetylcholine in order to signal muscles to move. In patients with myasthenia gravis, the body’s immune system attacks the receptors for acetylcholine. As a result, the muscles do not properly receive a signal from the nerves, and they become weak.

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How is ocular myasthenia gravis different from generalized myasthenia gravis?

Ocular myasthenia gravis only affects the muscles that move the eyes and eyelids. The symptoms of ocular myasthenia gravis include double vision (seeing two images instead of one), trouble focusing, and drooping eyelids. On the other hand, generalized myasthenia gravis affects muscles throughout the body. In addition to visual symptoms, generalized myasthenia gravis may cause trouble speaking, trouble swallowing, and weakness in the arms or legs.

The visual symptoms of myasthenia gravis include:
- Double vision
- Trouble focusing
- Drooping eyelids

The severity of the symptoms can fluctuate and be worse with fatigue.
Why does ocular myasthenia gravis typically cause double vision?

The brain finely controls the eye muscles in order to keep the eyes aligned properly. Weakness of the eye muscles leads to misalignment of the eyes, which causes the eyes to perceive the same object in two different locations. Double vision is a common symptom of myasthenia gravis because this condition very frequently affects the strength of the eye muscles.

How often does myasthenia gravis only affect the eyes?

For about one half of patients with myasthenia gravis, the first symptoms are visual. About 15% of these patients will remain only having visual symptoms, even years after their diagnosis. In the other 85% of patients, however, symptoms of weakness will develop in another part of the body, usually within the next three years. At that time, these patients are considered to have generalized myasthenia gravis.
How is ocular myasthenia gravis diagnosed?

Your doctor will begin by asking about your symptoms and performing a physical examination. It will be important to carefully test the eyelids and eye movements, and to see what happens to the muscles when they are fatigued and when they have rested.

Your doctor may request additional tests to confirm a diagnosis of myasthenia gravis. A blood test can check if the immune system is blocking the signals that travel from the nerves to the muscles. In patients with ocular myasthenia gravis (who have visual symptoms only) this blood test has lower accuracy, so a negative result might require additional testing.

One important test to help diagnose myasthenia gravis is called a nerve conduction study/electromyogram (sometimes called “EMG” for short). In part of this test, a small electrical stimulation is delivered to a nerve, and the responses are measured from a muscle. The most accurate test to diagnose myasthenia gravis is called a single fiber electromyogram (SFEMG). An SFEMG test involves very small needles that can measure responses from two nearby muscle fibers. Both the EMG and SFEMG tests can reveal a pattern of responses that suggests myasthenia gravis.

To diagnose ocular myasthenia gravis, a doctor might:

- Test the eyelids and eye movements
- Run a blood test
- Perform special studies (EMG or SFEMG) to measure the electrical responses in nerves and muscles
What are the treatments for ocular myasthenia gravis?

One of the medications that is commonly used to treat myasthenia gravis is called pyridostigmine (Mestinon). This medication helps build higher levels of the chemical acetylcholine, which is the signal that tells a muscle to move. The side effects of pyridostigmine can include diarrhea, abdominal cramps, nausea, and vomiting. Another medication called glycopyrrolate can be used to reduce these effects.

Corticosteroid medications, such as prednisone, can also be used to help patients with myasthenia gravis. These medicines can be very effective at controlling the immune response that causes myasthenia. Unfortunately, long-term use of steroids is also associated with side effects that include osteoporosis (weakening of bones), diabetes, high blood pressure, sleep disturbance, and emotional changes. In some cases, other medications can be used to help control the immune system and lower the dosage of prednisone. These medications include azathioprine (Imuran), cyclosporine, and mycophenolate mofetil (Cellcept).

If myasthenia gravis is diagnosed, a CT scan of the chest should be performed in order to look for a type of benign tumor called a thymoma. Many experts believe that if this tumor is present, removing it can improve the symptoms of myasthenia. Thymomas are present more often in patients with generalized myasthenia gravis compared to those with ocular myasthenia gravis.

Treatments for ocular myasthenia gravis include:

- Pyridostigmine (Mestinon) to build acetylcholine levels and glycopyrrolate to reduce the side effects
- Corticosteroids such as prednisone to control the immune response
- If there is a thymoma (tumor), removing it may improve symptoms
How can double vision be treated?

Double vision that results from myasthenia gravis is most often treated by blocking the vision from one eye. Either an eye patch can be worn or scotch tape can be placed over one lens in the eyeglasses. If one eyelid is very droopy and blocks the vision in that eye, the double vision will not require a separate treatment. Usually special glasses with prisms are not helpful because of the considerable fluctuation in the amount of double vision caused by myasthenia gravis.

Pyridostigmine (Mestinon) is a medication that reverses the symptoms of myasthenia gravis by boosting acetylcholine levels.
Patients with myasthenia gravis should avoid medications including:

- Specific antibiotics (aminoglycosides and quinolones)
- Beta-blockers
- Calcium channel blockers
- Phenytoin (Dilantin)

What medications should be avoided?

Certain medications can worsen the symptoms of myasthenia and should be avoided. These medications include specific antibiotics (called aminoglycosides and quinolones), beta-blockers, calcium channel blockers, and phenytoin (Dilantin). If a patient with myasthenia gravis requires a surgical procedure, the anesthesiologist should be aware in order to use the proper medications.

Where can I get more information on ocular myasthenia gravis?

For more information on ocular myasthenia gravis, consider the following sources:

Myasthenia Gravis Foundation of America
http://www.myasthenia.org/WhatisMG.aspx

National Institutes of Health

North American Neuro-Ophthalmology Society
http://www.nanosweb.org/i4a/pages/index.cfm?pageID=3286

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