



## Myasthenia Gravis

Your doctor thinks that you have Myasthenia Gravis (MG). This is an autoimmune condition where the body's immune system has damaged receptors on your muscles. This results in muscle weakness as receptors are necessary for the muscles to know when to contract. If this involves the muscles of the eyelid, it can result in lid droop (ptosis). If it involves the muscles of eye movement, it can result in double vision. The double vision and lid droop may vary, being worse when tired or later in the day.

### Anatomy:

When you want to move a particular muscle, the brain sends a signal via the nerves that go to the muscle. When this signal reaches the end of the nerve, it causes release of a chemical neuro-transmitter that diffuses across a narrow gap (the synapse) between the nerve and muscle. On the muscle side of the synapse, there are chemical receptors that are waiting to detect this neuro-transmitter. When they sense the presence of the neuro-transmitter, they begin the process that results in the muscle contracting. In the case of a patient with MG, when the neuro-transmitter is released and diffuses across the synapse, it does not produce normal muscular muscle contraction. This is because the normal receptors have been blocked or eliminated by antibodies produced by the body's immune system. Repeat attempts at moving the muscle will result in a gradual increase in weakness. Hence, muscles that are used most frequently are most likely to be weak. This is particularly true of the muscles that move the eye, resulting in ocular misalignment (double vision), or the muscles that lift up the lid, resulting in lid droop. Muscles anywhere within the body may be affected.

### Physiology:

The reason for the body's immune system's attack on the muscles is unclear. Patients with MG may have other autoimmune problems (such as thyroid orbitopathy). Like other autoimmune diseases, MG often spontaneously gets better but may come back. Over time, there may be some lessening effect, but MG produces variable disease that may last indefinitely or at least for years.

### Symptoms:

The most common symptoms of MG relate to weakness of the muscles that lift up the lid (ptosis) or move the eyes (double vision). MG can affect muscles anywhere in the body including those of swallowing or even breathing. Shortness of breath or difficulty swallowing may be very serious symptoms of MG and must be brought to your doctor's attention immediately. MG does not produce pain or numbness. If pain is present there must be something else going on and you need to tell your doctor.

### Signs:

The most common sign seen by ophthalmologists are lid droop



and eyes that don't work together. The eye movement problems may be difficult to detect and often patients with double vision related to MG have had it years before a diagnosis is made. It is not unusual for previous evaluations to have been normal.

#### Diagnosis:

MG can be confirmed by a biopsy of muscles and studies that demonstrate decreased receptors. In most cases, the diagnosis can be made without this procedure. A blood test may make the diagnosis, but about half of patients with MG restricted to the eye muscles may not have a positive test. Thus the finding of a negative blood test does not exclude MG. If Tensilon (a substance that decreases the turnover of the neuro-transmitter) is injected into a vein a patient with MG may demonstrate rapid improvement in the apparently weak muscle. Immediate elevation of the eyelid or improvement in eye movement can diagnose MG. A Tensilon test may be difficult to interpret if there are only mild findings. The sleep test or ice test depends on improved strength in rested muscles. If the patient is examined immediately upon awakening from rest, the findings of improved motility or resolution of lid droop may strongly suggest a diagnosis of MG. It is also possible to do studies of conduction between the nerve and muscle. This test, an EMG (electromyogram), has a relatively high sensitivity for the diagnosis of myasthenia. This requires sophisticated equipment and may be done only infrequently.

#### Prognosis:

As with other auto-immune conditions, MG tends to come and go. Symptoms may suddenly become worse. This can be precipitated by the use of certain antibiotics and other drugs. Once the diagnosis of MG is made, it is important that patients inform all their physicians, particularly if they are ever seen in an Emergency Room.

#### Treatment:

MG can be treated by medications that decrease the turnover of normal neuro-muscular transmitters. These may be taken in pill form and can be effective in improving weak muscles. Unfortunately, often this will not relieve lid droop or problems with the eye movements that result in double vision. Occasionally surgery to remove the thymus gland may be effective in treatment of MG and it is common to check for thymus enlargement with a chest CT scan. Steroid therapy (prednisone) may also be effective in improving muscle function. As steroids need to be used for long term, it is important to consider whether or not the benefits are outweighed by the risks of side effects associated with steroids. Occasionally other medications may help decrease the need for prednisone. This needs to be discussed with your physician.

#### Most frequently asked questions:

##### *How did I get Myasthenia?*

As with other autoimmune diseases, the reason for development of MG remains unclear. It is possible that some external challenge (viruses, etc.) may have caused the immune system to start acting in this fashion. At this time, however, we do not have full understanding of why certain patients develop MG.

##### *Is there a cure for Myasthenia?*

At this time there is no known cure for the autoimmune condition MG. Medical therapy may be effective in improving muscular function but needs to be tailored to the particular activity of the disease process. Drugs that influence the immune system response may also be helpful.

##### *What are my chances of developing weakness in muscles other than my eye muscles?*

MG may seem to involve only certain muscles, especially the muscles that move the eye and lift the lid. It has been felt that when MG fails to cause general muscle weakness over 2 years or more that the chances of developing weakness in other non-ocular muscles in the future is low. This is probably true but not a guarantee that systemic weakness will not occur. It remains very important, that if problems with breathing or swallowing ever develop, it needs to be brought to the attention of your physician immediately.