Understanding Myasthenia Gravis

Jerry, diagnosed in 2002.
What Is Myasthenia Gravis?

Myasthenia gravis (MG) is a neuromuscular disorder that causes muscle weakness. It affects muscles that a person can usually control consciously. Muscles most commonly affected are those controlling the eyelids, eye movement, and breathing and swallowing, as well as the facial and shoulder muscles. The weakness tends to temporarily worsen with activity and improve with rest. MG is an autoimmune disorder. This means the body’s immune system mistakenly attacks the connection between the nerve and muscle. In MG, the muscle cells have problems responding to the nerve impulses that normally signal them to contract, and this results in weakness.

The disease usually does not reduce life expectancy. Most people with MG manage their symptoms and lead active lives. MG affects people of all ages and ethnic groups. Women are most often affected in their 20s and 30s. Men usually develop MG in late middle age.

Transmission of Nerve Impulses to Muscles

![Diagram of nerve cell and muscle with receptors and acetylcholine](image)
What Causes Myasthenia Gravis?

MG is caused by a problem with nerve impulses getting to muscles. Normally when impulses travel down the nerve, the nerve endings release a protein called acetylcholine. Acetylcholine is like a key that fits into the acetylcholine receptor, which is like a lock. With the proper fit, the door is opened and a signal is sent to the muscle to do its job. Antibodies are proteins produced by the body in response to harmful substances. In MG, the body mistakenly makes antibodies to the acetylcholine receptor. Acetylcholine receptor antibodies fit into the receptor and cause it to be destroyed so the door does not open and no signal is received by the muscle. This leads to muscle weakness.

It is thought that the thymus gland may instruct the immune system to produce the antibodies. The thymus gland is in the chest. In some people with MG, the thymus becomes enlarged. In others, tumors of the thymus may develop.

In rare cases, children develop MG symptoms due to a genetic defect. In these children, acetylcholine antibodies cannot be detected in the blood.

Women with MG who are planning pregnancies should talk to their physicians. Temporary MG occurs in about 15 percent of infants born to women with MG due to transfer of antibodies across the placenta. The condition lasts for only a few weeks, but it needs to be recognized. These infants often need to be treated in an intensive care unit until the weakness is gone.
What Are the Symptoms?
The symptoms of MG vary from one person to the next. For some, only the muscles that control eye movements and the eyelids are affected. In others, the muscles that control swallowing and speech are affected. Some muscles are weaker than others. In some people, breathing can be affected. This is due to weakness of the throat or diaphragm muscles. In a few cases, weakness can cause respiratory failure that requires emergency medical support. In contrast to many other diseases producing weakness, the legs are less commonly involved in MG.

How Is Myasthenia Gravis Diagnosed?
Many disorders can cause weakness. The diagnosis of MG should be made by a neurologist. The evaluation may include:

- Medical and neurologic evaluation
- Blood tests to check for antibodies
- Blood tests or other studies to rule out other causes of weakness
- Imaging scans
- Electrical tests of nerve and muscle function (electromyography and nerve conduction studies)
- Ice pack test to improve strength of the eyelid

What Are the Treatment Options?
MG does not yet have a cure, but effective treatments are available. It can take time to find the right combination of treatments for each person. In up to 20 percent of people symptoms may improve or disappear for up to a year or more. Occasionally the disease may disappear permanently, although this is rare. No one knows why these remissions occur.

Treatments include:

- **Medication**
  - Drugs to suppress the immune system and decrease the autoantibodies
  - Drugs that slow the breakdown of acetylcholine and prolong its action at the nerve-muscle connection
- **Surgery**: The thymus gland should be removed if the thymus is enlarged on imaging as it could indicate a tumor. It is still not clear whether removal of a thymus that appears normal on imaging is a useful treatment.
Neurologists are medical doctors who specialize in disorders of the brain and nerves. They are the specialists who most often diagnose and treat people with myasthenia gravis.

- **Plasma exchange**: In plasma exchange, abnormal antibodies are removed from the blood. Then red blood cells are returned in artificial plasma. Antibody removal in this way produces temporary improvement in the majority of patients but also requires some form of immune suppression so that the antibodies do not rebound.

- **Intravenous immunoglobulin (IVIG)**: Treatment involves administering purified antibodies. These temporarily modify the immune system and provide the body with normal antibodies from donated blood.

### Living with Myasthenia Gravis

With treatment, the outlook for most people with MG is bright. You can have a full, productive life.

Learning to manage the symptoms of MG can make it easier. MG support groups can be a source of help, comfort, and information. Counseling can help in coping with the emotional aspects of MG.

People with MG need to plan their activities to conserve their energy:
- Schedule regular rest periods during the day
- Delegate tasks to other family members
- Learn to manage and reduce your stress
- If your swallowing is affected, plan meals at times when your muscle strength is greater

### Partnering with Your Neurologist

To provide the best care, your neurologist needs to know all about your symptoms and medical history. Likewise, you need to get answers to your questions. Keeping a notebook about your condition and bringing a few well-organized questions to your appointments can be helpful.

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For Family and Friends

MG can take a toll on the family, too. If you are caring for a family member or friend with MG, take care of yourself, as well. Avoid the feeling that you have to do it all yourself. Get help from family, friends, and professionals. There are many support groups for caregivers. See Resources for organizations to contact.

Help Us Cure Brain Disease

Make a Donation to Research
The American Brain Foundation supports vital research and education to discover causes, improved treatments, and cures for brain and other nervous system diseases. To learn more or to make a donation to support research, visit www.CureBrainDisease.org.

Make Your Voice Heard
To keep research advancing toward future cures and treatments for brain disease, it is important for people affected by neurologic disorders to advocate for more research funding. Contact your members of Congress and ask them to support neurology research by increasing funding for the National Institutes of Health (NIH). Look up your Congressional representatives at www.senate.gov and www.house.gov. Your voice can make a difference.

Take Part in Research
People are needed for clinical trials that can help find new treatments for neurologic disorders. Clinical trials are research studies. They help ensure that new drugs are both safe and effective. Ask your neurologist how to volunteer for a clinical trial. You can also find trials through patient organizations or the American Academy of Neurology website at www.aan.com/view/clinicaltrials.
Resources

American Academy of Neurology
www.aan.com/patients
(800) 879-1960
The American Academy of Neurology website for patients and caregivers offers a wealth of articles, information about events and resources, and links to support groups, clinical trial information, and more.

Neurology Now® magazine
www.neurologynow.com
(800) 879-1960
Free magazine for patients and caregivers, courtesy of the American Academy of Neurology. Stories about people living with neurologic disorders, the latest information on resources and treatments, and more.

American Autoimmune Related Diseases Association
www.aarda.org
(586) 776-3900

Muscular Dystrophy Association
www.mdausa.org
(800) 572-1717

Myasthenia Gravis Foundation of America
www.myasthenia.org
(800) 541-5454
The American Academy of Neurology, an association of 25,000 neurologists and neuroscience professionals, is dedicated to promoting the highest quality patient-centered neurologic care. For more information about the American Academy of Neurology, visit www.aan.com.