



Frequently Asked Questions about Myasthenia Gravis

What is myasthenia gravis (MG)?

Myasthenia gravis is an *autoimmune* disease that affects the transmission of signals from nerves to muscles. The name myasthenia gravis comes from Greek and Latin words meaning "grave muscle weakness." Today, however, most cases of MG are not as "grave" as the name implies. In fact, most people with MG can expect to live normal or nearly normal lives.

The hallmark of MG is muscle weakness that increases during activity and improves after rest. MG often involves muscles that control eye and eyelid movement, facial expression, chewing, talking, and swallowing. The muscles that control breathing and neck and limb movements may also be affected.

The thymus gland, part of the immune system, is abnormal in most MG cases. Some people with MG have *benign* (noncancerous) tumors of the thymus gland called *thymomas*.

Some drugs can trigger or worsen MG symptoms.

What causes MG?

MG is caused by a defect in the transmission of nerve signals to muscles. Normally, nerve endings release a substance called *acetylcholine* that binds or attaches to *receptors* on the muscle. This leads to muscle contractions. In MG, the body's own immune system produces antibodies that block this transmission.

Who gets myasthenia gravis?

Estimates of the number of people affected by MG vary, ranging from five to 14 people per 100,000.

MG occurs in all ethnic groups and both genders. It most commonly affects young adult women (under 40) and older men (over 60), but it can occur at any age. Children sometimes develop MG.

MG is not directly inherited nor is it contagious. Sometimes the disease may occur in more than one member of the same family. If a woman with MG becomes pregnant, sometimes the baby acquires antibodies from the mother and has MG symptoms for a few weeks or months after birth. This is called *neonatal myasthenia*, and the symptoms can be treated.

In rare cases, myasthenia is caused by a defective gene and appears in infants born to non-myasthenic mothers. This type is called *congenital myasthenia*.

What is the role of the thymus gland in MG?

The thymus gland, found in the upper chest area beneath the breastbone, is a part of the body's normal immune system. In most adults with MG, the thymus gland is abnormal. Some people with MG develop thymomas or tumors on the thymus gland. Generally thymomas are benign, but they can become *malignant* (cancerous). The relationship between the thymus gland and MG is not yet fully understood.

project of the U.S. Department of Health and Human Services Office on Women's Health

What are the signs and symptoms of MG?

The muscles that control eye and eyelid movement, facial expression, and swallowing are most often affected. The onset of the disorder may be sudden. Symptoms often are not immediately recognized as MG.

In most cases, the first noticeable symptom is weakness of the eye muscles. In others, difficulty in swallowing and slurred speech may be the first signs. While rare, first signs of MG can also include difficulty with breathing. The degree of muscle weakness involved in MG varies greatly among persons with this disease. Symptoms, which vary in type and severity, may include:

- Drooping of one or both eyelids (ptosis)
- Blurred or double vision (*diplopia*) due to weakness of the muscles that control eye movements
- Unstable or waddling gait
- Weakness in arms, hands, fingers, legs, and neck
- Change in facial expression
- Difficulty in swallowing and shortness of breath
- Impaired speech (dysarthria)
- Shortness of breath

How is MG diagnosed?

Unfortunately, a delay in diagnosis of one or two years is not unusual in cases of MG. Weakness is a common symptom of many other disorders. The diagnosis is often missed in people who have mild weakness or in those whose weakness is restricted to only a few muscles.

The first steps of diagnosing MG include a review of the person's medical history and physical and neurological exams. If the doctor suspects MG, several tests are available to confirm the diagnosis.

- Antibody blood test. A special blood test can detect the antibodies that prevent nerves from signaling to muscles. While most people with MG have abnormally high levels of these antibodies, some individuals (about 10 percent) can actually test negative for antibodies. And, antibodies may not be detected if only eye muscles are affected.
- Edrophonium test. When this drug is injected, the weak eye muscles of people with MG will briefly get stronger.
- Nerve conduction test/repetitive stimulation. This is a test of specific muscle fatigue by repetitive nerve stimulation.
- Single fiber *electromyography* (EMG). In this test, pairs of single muscle fibers are stimulated by electrical impulses. It can detect impaired nerve-to-muscle transmission.

www.4woman.gov -- 800-994-WOMAN (9662) -- 888-220-5446 (TDD)

<u> 4 woman.gov – The National Women's Health Information Center</u>

project of the U.S. Department of Health and Human Services Office on Women's Health

• *Computed tomography* (CT) or *magnetic resonance imaging* (MRI). These tests can help identify an abnormal thymus gland or a thymus gland tumor.

What is the treatment for MG?

Today, MG can be controlled. There are several therapies available to help reduce muscle weakness. Most persons with MG have good results from treatment. In some people MG, like many other autoimmune diseases, may go into *remission* (a period of time without symptoms) and muscle weakness may disappear completely.

Remission or improvement can occur without treatment in some cases. According to the Muscular Dystrophy Association, up to 20 percent of person with MG may have complete remission of symptoms without any treatment, and another 20 percent may improve without treatment. These spontaneous improvements are more likely to occur during early stages of MG.

Treatment of MG may include:

- Medications. Drugs used include *cholinesterase* inhibitors such as *neostigmine* and *pyridostigmine*. These drugs help improve nerve signals to muscles and increase muscle strength. *Immunosuppressive* drugs such as *prednisone*, *cyclosporine*, and *azathioprine* may also be used to suppress the production of abnormal antibodies. They must be used with careful medical followup because they can be associated with major side effects.
- *Thymectomy*, the surgical removal of the thymus gland (which is abnormal in most persons with MG). This surgery is done for persons with MG who have tumors, as well as for individuals without tumors. It improves symptoms in more than half of individuals without tumors. It may cure some people with MG, possibly by re-balancing the immune system.

Other therapies sometimes used to treat MG during especially difficult periods of weakness include:

- *Plasmapheresis* or plasma exchange. This is a procedure that removes abnormal antibodies from the blood.
- High-dose intravenous immune globulin. This treatment temporarily interferes with the ability of the immune system to damage the nerve muscle junction.

Treatment options for a person with MG depend on the severity of the weakness, which muscles are affected, and the person's age and other medical problems.

In a few cases, MG may cause severe weakness resulting in acute respiratory failure. But most people can expect to lead normal or nearly normal lives.

What is a myasthenic crisis?

A myasthenic crisis occurs when weakness affects the muscles that control breathing. This can create a medical emergency requiring a respirator to help the person breathe or measures to prevent a person from taking in, or aspirating, too much air into their lungs. In individuals whose respiratory muscles are weak, infection, fever, a reaction to medication, or emotional stress can trigger a crisis.

www.4woman.gov -- 800-994-WOMAN (9662) -- 888-220-5446 (TDD)

<u> 4 woman.gov – The National Women's Health Information Center</u>

project of the U.S. Department of Health and Human Services Office on Women's Health

How can I help take care of myself if I have MG?

You can follow a few simple steps to help cope with the condition in your daily life. Plenty of rest and a well balanced, potassium-rich diet can help ease fatigue. Good sources of potassium include oranges, orange juice, and bananas. It is important to avoid overexertion, and if necessary, to rest the eyes or to lie down briefly a few times a day.

Is MG associated with other conditions?

Since it is an autoimmune disease, it may occur in combination with other autoimmune conditions such as *rheumatoid arthritis*, *Sjorgrens syndrome*, *lupus*, *pernicious anemia*, or *autoimmune thyroiditis*.

For more information...

You can find out more information about Myasthenia Gravis by contacting the National Women's Health Information Center at (800) 994-9662 or the following organizations:

National Institute of Neurological Disorders and Stroke

Phone Number(s): (800) 352-9424 Internet Address: <u>http://www.ninds.nih.gov</u>

Myasthenia Gravis Foundation

Phone Number(s): (800) 541-5454 Internet Address: <u>http://www.myasthenia.org</u>

American Autoimmune Related Diseases Association

Phone Number(s): (810) 776-3900, (800) 598-4668 Literature Requests Internet Address: <u>http://www.aarda.org</u>

This fact sheet was abstracted primarily from publications of the National Institute of Neurological Disorders and Stroke (NINDS).

All material contained in the FAQs is free of copyright restrictions, and may be copied, reproduced, or duplicated without permission of the Office on Women's Health in the Department of Health and Human Services; citation of the source is appreciated.

This FAQ reviewed by Al P. Kerza-Kwiatecki, Ph.D., National Institute of Neurological Disorders and Stroke, National Institutes of Health July 2002

www.4woman.gov -- 800-994-WOMAN (9662) -- 888-220-5446 (TDD)