

WHAT IS Myasthenia Gravis?

Taken from the MGF of Illinois Web site

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What is MG?

Myasthenia gravis (MG) is an autoimmune disease. With MG, the immune system targets the communication point between the nerve and muscle (called the “neuromuscular junction”). MG causes weakness in voluntary muscles; this weakness worsens with activity and improves with rest.

MG’s primary symptoms fluctuate, vary in severity and occur in many combinations. Symptoms may include droopy eyelids(s); double or blurred vision; weak arms, hands, neck, face or legs; difficulty chewing, smiling, swallowing or talking; excessive fatigue; difficult breathing or shallow respirations.

The name myasthenia gravis comes from Greek and Latin words meaning “grave muscle weakness.” This reflects the situation for patients with MG before the discovery of many of the current treatments. Today, with appropriate therapy, most people with MG can expect to live normal or nearly normal lives. Also, sustained clinical remissions, although rare (10-15% of cases), can happen at any time.

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What Causes MG?

MG is caused by a defect in the way nerves transmit signals to muscles. Normally, impulses travel down the nerve, and nerve endings release a substance called *acetylcholine*. Acetylcholine binds or attaches to receptors on the muscle and makes the muscle contract. The strength of the contraction depends on how much acetylcholine the muscles receive.

With MG, your immune system produces antibodies that block or destroy many of the receptor sites for acetylcholine in your muscles (the acetylcholine receptors). Because there are fewer receptor sites, the muscles get fewer nerve signals and become weak.

In another form of MG, your immune system produces antibodies to the MuSK (muscle specific tyrosine kinase) protein. This also interferes with the nerve-to-muscle communication and causes muscle weakness.

Although not fully understood, doctors believe the *thymus gland* likely plays a role in starting the abnormal immune response in some MG patients. This gland, which is large in infancy, is involved in the education and development of your immune system. Usually the gland shrinks and becomes dormant in adults, turning into fatty tissue in the upper chest behind the breastbone. With patients whose MG symptoms begin before age 40, the thymus gland is

generally larger than normal, and may be responsible for triggering the immune attack on the neuromuscular junction. Some people with MG develop “thymomas” or tumors on the thymus gland. Generally thymomas are benign, but in rare cases they can become malignant. When a thymoma is discovered, surgery to remove it should be performed.

Physicians aren't sure what triggers the onset of MG. They suspect there may be a combination of factors. For example, if you have an abnormal thymus gland or genetic predisposition, then an illness or other event that stimulates the immune system could trigger myasthenia.

This information was abstracted from the *Myasthenia Gravis Fact Sheet* published by the National Institute of Neurological Disorders (2007, December 13) and retrieved March, 27, 2008 from http://www.ninds.nih.gov/disorders/myasthenia_gravis. Additional medical references were used as well. Reviewed by the MGF of Illinois Medical Advisory Board, September 2008.

Who Gets MG?

It is difficult to determine how many people actually have MG because the disease is often undiagnosed or misdiagnosed. However, it's estimated that there are two to seven people in every 10,000 that have myasthenia gravis (Muscular Dystrophy Association, “Facts About Myasthenia Gravis,” May 2006). MG occurs in all ethnic groups throughout the world, and in both genders. According to the National Institute of Neurological Disorders and Stroke, it most commonly affects women under age 40, and men over age 60, but it can occur at any age.

MG is not thought to be directly inherited. It is not contagious. Myasthenia gravis is more common in families with other autoimmune diseases. 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 very rare cases, a form of myasthenia can be caused by a defective gene and appears in infants born to non-myasthenic mothers. This form of myasthenia is called *congenital myasthenic syndrome*; it involves problems with the ACh receptors, not the immune system.

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Symptoms of MG

Myasthenia gravis can affect any of the muscles that you control voluntarily. It can affect muscles of the face, hands, eyes, arms and legs and those muscles involved in chewing, swallowing and talking. Muscles that control breathing and neck movement also can be affected.

MG does not affect involuntary muscles such as the heart, smooth muscles of the gut, blood vessels, and uterus.

For most people, the first noticeable symptom is weakness of the eye muscles causing drooping eyelids or double vision. In others, difficulty in swallowing and slurred speech may be the first signs. The onset of the disease is usually gradual, but may be sudden. Symptoms may come

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and go over time. Symptoms often are not immediately recognized as MG, especially if they are subtle or variable.

Symptoms, which vary in type, severity and combination, may include:

- Drooping of one or both eyelids
- Double or blurred vision
- Weakness in arms, hands, neck, face or legs
- Difficulty in chewing, smiling, swallowing or talking
- Excessive fatigue in exercised muscle groups
- Difficult breathing or shallow respiration

Most individuals do not develop all of the symptoms.

Myasthenia gravis is often called the “snowflake disease” because it differs so much from person to person. The degree of muscle weakness and the muscles that are affected vary greatly from patient to patient and from time to time.

While weakness in the eye muscles is the most common initial symptom in MG, in some patients weakness remains limited to the eyes for entire course of the disease. These patients have *ocular MG*. Others have *generalized MG* where symptoms are present throughout the body, usually including the eyes. For instance, it may be hard to hold up an arm to comb your hair or shave or put on make-up. With a weakened grip, it may be difficult to open jars. Weak hips may make it difficult to get out of deep chairs or the bathtub. Legs may tire when climbing stairs.

People with MG are more likely to suffer from another autoimmune disease as well. These can include thyroid disease, lupus, rheumatoid arthritis and diabetes. With thyroid disease, an episode of hypothyroidism may trigger a flare-up of MG weakness.

What is an MG Crisis?

If a person’s ability to breathe, cough, or protect their airway becomes insufficient, it’s called a *myasthenic crisis*. These patients need mechanical breathing assistance in a hospital for a period of time until their strength improves. While most myasthenics never experience a crisis, those who have trouble swallowing and talking are the ones most likely also to have trouble breathing. Before a crisis happens usually there are progressive warning signs that swallowing, talking, and breathing are becoming compromised.

This information was abstracted from these sources:

- National Institute of Neurological Disorders (2007, December 13). *Myasthenia Gravis Fact Sheet*. Retrieved March, 27, 2008 from http://www.ninds.nih.gov/disorders/myasthenia_gravis
 - Myasthenia Gravis Foundation of Illinois, *Conquer* newsletter, May 2008
- Reviewed by the MGF of Illinois Medical Advisory Board, September 2008.

Factors that Worsen MG

Some factors can make myasthenia worse:

- fatigue, insufficient sleep
- stress, anxiety
- illness
- overexertion, repetitive motion
- pain
- hot foods or beverages (affects mouth and throat)
- some medications, including beta blockers, calcium channel blockers, and some antibiotics

- sudden fear, extreme anger
- depression
- extreme temperatures (hot or cold weather, hot showers or baths, sunbathing, saunas, hot tubs)
- humidity
- sunlight or bright lights (affects eyes)
- alcoholic beverages
- quinine or tonic water
- low potassium levels or low thyroid levels
- exposure to some chemicals, including some household cleaners, insecticides and pet flea sprays
- chemical lawn treatments

Infections and respiratory illnesses can produce increased weakness that lasts for a while after the illness is gone. The stress of surgery can make MG temporarily worse. The disease may intensify during certain times of a woman's menstrual cycle.

A list of drugs that can make your MG worse has been compiled by the University of Illinois at Chicago (<https://www.uic.edu/pharmacy/services/di/myasthenia.htm>). Also, be sure to check with your primary MG doctor before taking any new medication, even over-the-counter drugs.

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Diagnosis

Myasthenia gravis can be difficult to diagnose because weakness is a common symptom of many disorders. Add to this the fact that symptoms may be vague, fluctuate or only affect certain muscles. And MG doesn't "perform" on demand; the eyelid that droops at 7 p.m. may not show for a 9 a.m. doctor appointment. Identification may be complicated further when more than one autoimmune disorder is involved. It's not unusual for a diagnosis of MG to be delayed.

These steps may be taken to confirm a diagnosis of MG:

- **Neurological exam.** This may include testing your reflexes, muscle strength, muscle tone, senses of touch and sight, gait, posture, coordination, balance and mental skills. Impaired eye movement or muscle weakness may prompt a doctor to evaluate further.
- **Blood tests to measure myasthenic antibodies in the blood.** About 80% of all MG patients have elevated levels of AChR antibodies that block or destroy acetylcholine receptor sites on the muscles. However, blood tests may not be conclusive, particularly in patients with mild cases of MG or with ocular MG.
- **Tensilon® test.** A short-acting drug called edrophonium chloride (Tensilon®) is given intravenously. If weakness, especially in the eye muscles, briefly and temporarily improves, it indicates you may have myasthenia gravis. The drug does this by blocking an enzyme that breaks down acetylcholine, the chemical that transmits signals from nerve endings to muscles. A trial use of oral pyridostigmine bromide (Mestinon) is an alternative approach.
- **Nerve conduction studies/repetitive nerve stimulation.** A small electrical impulse is applied to your skin. This stimulates your nerves to test the strength of your muscle contraction. Muscle fibers in patients with MG fatigue easily and don't bounce back from repeated stimulation as well as those of a healthy person.
- **Single fiber electromyography (EMG).** A thin-needle electrode is inserted into one of your muscles to help measure patterns of electrical activity in your muscle at rest and with slight muscle contraction. The test can detect defective nerve-to-muscle communication.
- **Imaging.** Computed tomography (CT) or magnetic resonance imaging (MRI) are often done to identify an abnormal thymus gland or a thymus gland tumor (thymoma).

There is no one foolproof method of diagnosis for MG. Sometimes test results are inconclusive, even when your examination and story seem to point to a diagnosis of the disease. If this happens, it may be appropriate to track symptoms over time until your diagnosis can be clarified.

On occasion, some testing is done to rule out other disorders.
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Treatment and Prognosis

With [treatment \(LINK: TREATMENT SECTION\)](#), people with MG have a normal life expectancy. Some combination of medication, thymectomy, and other therapies enables most myasthenics to lead normal or near normal lives. Sometimes people experience remission. However, for some people quality of life is affected significantly—either by severity of the disease or severity of side effects from the medication.

Generally, those who are quickly diagnosed and receive effective treatment have the best outcomes.

The first two or three years of illness are often when the full extent of MG symptoms emerge, and so can be the most difficult. During this time you may need to try a number of different therapies, to see which you tolerate best and which is most effective.

Myasthenia gravis generally doesn't get worse as you age. And for some, symptoms diminish over time.

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