

What is Myasthenia Gravis?

Myasthenia Gravis is a chronic neuromuscular disease deriving its name from Latin and Greek words meaning “grave muscle weakness.” The disease is characterized by abnormal weakness of voluntary muscles (those muscles controlled by will). This weakness increases with activity and decreases with periods of rest.

Myasthenia Gravis may affect an individual of any age or race including the newborn child. However, the disease is seen more frequently in the young adult female and in the older male. The role of heredity in MG is uncertain.

Myasthenia Gravis, or MG, may involve either a single muscle or a group of muscles. The muscles which control chewing, swallowing and eye movement are most often affected, followed by the muscles that control the arms and legs. The muscles used for breathing may also be affected. Weakness in these muscles may result in shortness of breath; an inability to take a deep breath; or difficulty coughing.

Those MG patients who experience severe difficulty breathing usually require hospitalization.

To understand why the myasthenic has muscles that are weak, we need to explore how muscles function.

Muscles are controlled by the nervous system. For a voluntary muscle to contract so that you can chew food, talk, breathe or walk, a message is sent from the brain along a nerve pathway to the nerve ending. The nerve ending is very close to the muscle, but does not touch the muscle. The gap between the nerve ending and the muscle is called the neuromuscular junction. The message that has been sent from the brain to the nerve ending causes the release of the chemical, acetylcholine in the nerve ending. The acetylcholine carries the message to a special place on the muscle called a receptor site. Each neuromuscular junction, has many receptor sites. When a sufficient number of receptor sites have been activated by acetylcholine, the muscle contracts.

With MG, the muscle weakness occurs because there is a reduction in the number of receptor sites at the neuromuscular junction. This destruction of receptor sites is due to an antibody. The origin of the antibody is unknown. Because an antibody is involved in the destruction of the receptor sites, MG is classified as an autoimmune disease. To understand what the term “autoimmune” means, it will help to know about the body’s defense system.

Behind the breastbone is a small gland called the thymus. Early in life the thymus gland is involved in the development of the immune system which enables the body to defend itself against illness. The exact role of the thymus in MG is unknown. The thymus gland produces certain types of cells that are a vital part of the immune system. These cells stimulate the production of antibodies, which recognize foreign invaders called antigens. Normally the antibodies destroy antigens before major illness occurs.

With an autoimmune disease (and there are many different kinds), the antibodies become confused. Instead of attacking a foreign invader, the confused antibodies start attacking the body they are meant to protect. MG causes these confused antibodies attack the receptor sites of the neuromuscular junction. We now know that there is an 80 – 90 % reduction in the acetylcholine receptor sites in the

muscle due to the action of confused antibodies. The rate of destruction of the receptor sites is greater than the replacement of the receptor sites. Acetylcholine, therefore, does not reach enough receptor sites to cause strong muscle contractions. This leads to muscle weakness.

When attempting to identify the source of muscle weakness, the patient will undergo a neurological examination that includes testing of muscle strength. If the doctor suspects that the patient might have Myasthenia Gravis, he or she may do an edrophonium, or Tensilon, test. Special medication is injected into the vein. Muscle strength will be tested before and after the medication is injected. An improvement in muscle strength provides strong support for a diagnosis of Myasthenia Gravis. Another test that may be done to confirm the diagnosis of MG involves a repeated stimulation of a nerve, resulting in a particular type of muscle response. A blood sample may also be drawn to determine the presence of the confused antibody. In 90% of MG patients, there is an elevation of this antibody.

While there is no known cure for Myasthenia Gravis, it can be treated in a variety of ways. These treatments include medication, plasmapheresis, and thymectomy.

Medication: The most frequently used treatment is medication. The drugs most commonly used today are anticholinesterase agents such as Mestinon, corticosteroid drugs such as Prednisone, and other immunosuppressive agents such as Imuran and CellCept.

Plasmapheresis: Plasmapheresis removes the confused antibodies from the plasma portion of the blood. The striking improvement in strength following plasmapheresis is short-lived due to the continuing formation of the antibodies; therefore, the process of plasmapheresis will have to be repeated.

Thymectomy: The surgical removal of the thymus gland is called a thymectomy. Thymectomy may lessen the severity of myasthenic symptoms. The degree to which the symptoms are lessened differs with each patient.

The outlook for Myasthenia Gravis patients varies. Treatments not completely halt the disease but can improve the symptoms. In some cases, the disease may go into remission in which case the symptoms disappear and no treatment is necessary. A doctor will determine which type of treatment is best for the patient. Treatment of Myasthenia Gravis requires very careful attention to the prescribed therapy. There must be a good working relationship between the patient and the health care team. Myasthenia Gravis, though not curable by any known method, is controllable, allowing most patients to lead nearly full and productive lives.