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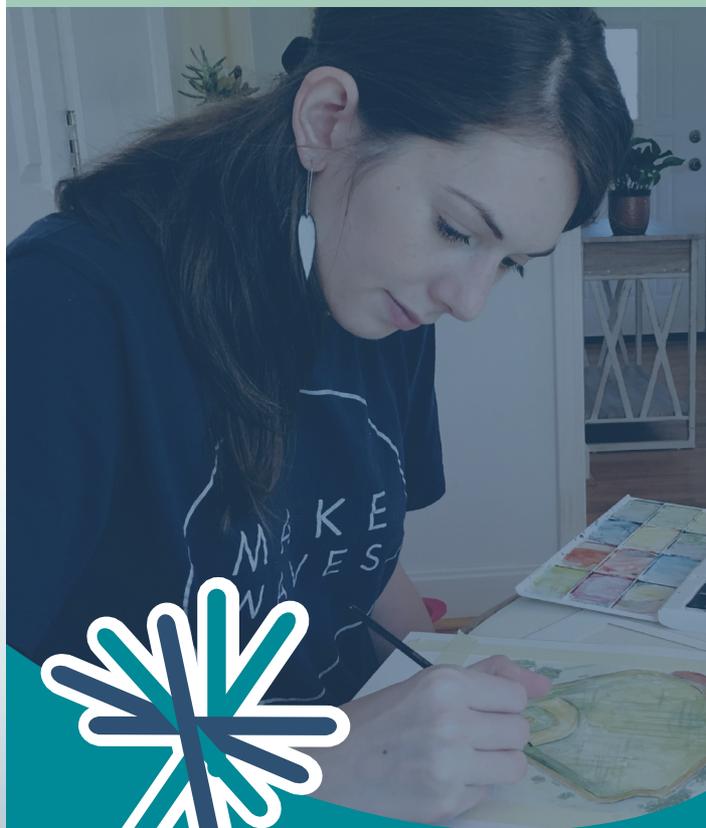
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Myasthenia Gravis (MG)

Myasthenia gravis comes from the Greek and Latin words meaning “grave muscular weakness.” The most common form of MG is a chronic autoimmune neuromuscular disorder that is characterized by fluctuating weakness of the voluntary muscle groups. The prevalence of MG in the United States is estimated to be about 20/100,000 population. However, MG is probably under diagnosed and the prevalence may be higher.

What Causes Autoimmune MG?

The voluntary muscles of the entire body are controlled by nerve impulses that arise in the brain. These nerve impulses travel down the nerves to the place where the nerves meet the muscle fibers. Nerve fibers do not actually connect with muscle fibers. There is a space between the nerve ending and muscle fiber; this space is called the neuromuscular junction.

When the nerve impulse originating in the brain arrives at the nerve ending, it releases a chemical called acetylcholine. Acetylcholine travels across the space to the muscle fiber side of the neuromuscular junction where it attaches to many receptor sites. The muscle contracts when enough

of the receptor sites have been activated by the acetylcholine. In MG, there is as much as an 80% reduction in the number of these receptor sites available. The reduction in the number of receptor sites is caused by antibodies that destroy or block the receptor site.

Antibodies are proteins that play an important role in the immune system. They are normally directed at foreign proteins called antigens that attack the body. Such foreign proteins include bacteria and viruses.

Antibodies help the body to protect itself from these foreign proteins. For reasons not well understood, the immune system of the person with MG makes antibodies against the receptor sites of the neuromuscular junction. Abnormal antibodies can be measured in the blood of many people with MG. There are currently three commercially available tests for the abnormal antibodies, the AChR (acetylcholine receptor), MuSK (muscle specific kinase) and low density lipoprotein related protein 4 (LRP4) antibodies. These antibodies destroy the receptor sites or make them unavailable. Muscle weakness occurs when acetylcholine cannot activate enough receptor sites at the neuromuscular junction.

Clinical Features and Symptoms

MG occurs in all races, both genders and at any age. MG is not directly inherited nor is it contagious. It does occasionally occur in more than one member of the same family.

MG may affect any muscle that is under voluntary control. Certain muscles are more frequently involved, and these include the ones that control eye movements, eyelids, chewing, swallowing, coughing and facial expression. Muscles that control breathing and movements of the arms and legs may also be affected. Weakness of the muscles needed for breathing may cause shortness of breath, difficulty taking a deep breath and coughing.

The muscle weakness of MG increases with continued or repetitive activity and improves after periods of rest. The muscles involved may vary greatly from one person to the next. In some people, weakness may be limited to the muscles controlling eye movements and the eyelids. This form of myasthenia is referred to as ocular MG. Generalized MG refers to those people with MG who have weakness involving muscles outside the eye region. In the most severe form of generalized MG, many of the voluntary muscles of the body are involved, including those needed for breathing. The degree and distribution of muscle weakness for many people falls in between these two extremes. When the weakness is severe and involves breathing, hospitalization is usually necessary.

Diagnosis

There are many disorders that cause weakness. In addition to a complete medical and neurological evaluation, a number of tests may be used to establish a diagnosis of MG. Blood tests for the abnormal antibodies can be performed to see if they are present.

Electromyography (EMG) studies can provide support for the diagnosis of MG when characteristic patterns are present. The edrophonium chloride (Tensilon®) test is performed by injecting this chemical into a vein. Improvement of strength immediately after the injection provides strong support for the diagnosis of MG. Sometimes all of these tests are negative or equivocal in someone whose story and examination still seem to point to a diagnosis of MG. The positive clinical findings should probably take precedence over negative confirmatory tests.

Treatment

There is no known cure for MG, but there are effective treatments that allow many—but not all—people with MG to lead full lives. Common treatments include medications, thymectomy, plasmapheresis (also called total plasma exchange and abbreviated as PLEX or TPE) and intravenous immunoglobulins (IVIg). Spontaneous improvement and even remission may occur without specific therapy.

Medications are most frequently used in treatment. Anticholinesterase agents (e.g., Mestinon®) allow acetylcholine to remain at the neuromuscular junction longer than usual so that more receptor sites can be activated. Corticosteroids (e.g., prednisone) and immunosuppressant agents (e.g., Imuran®, CellCept®) may be used to suppress the abnormal action of the immune system that occurs in MG. Intravenous immunoglobulins (IVIg) are also sometimes used to affect the function

or production of abnormal antibodies. Recently, monoclonal antibodies have been investigated for use in MG. Rituximab has been found to be an effective treatment for MuSK positive patients, and is currently being tried for those positive for AChR. In 2016, a new treatment for MG called Soliris (eculizimab) was approved to treat people with MG who are AChR positive and have not responded well to other therapies.

Thymectomy (surgical removal of the thymus gland) is another treatment used in some people with MG. The thymus gland lies behind the breastbone and is an important part of the immune system. When there is a tumor of the thymus gland (present in 10-15% of patients diagnosed with MG), it is nearly always removed because of the risk of malignancy. Thymectomy frequently lessens the severity of the MG weakness after some months. In some people, the weakness may completely disappear. The degree to which the thymectomy helps varies with each person.

Plasmapheresis, or plasma exchange (PLEX), may also be useful in the treatment of MG. This procedure removes the abnormal antibodies from the plasma portion of the blood. The improvement in muscle strength may be striking, but is usually short-lived, since production of the abnormal antibodies continues. When plasmapheresis is used, it may require repeated exchanges. Plasma exchange may be especially useful during severe MG weakness or prior to surgery.

Treatment decisions are based on knowledge of the natural history of MG in

each patient and the predicted response to a specific form of therapy. Treatment goals are individualized according to the severity of the MG weakness, the person's age and sex, and the degree of impairment.

Prognosis

The current treatments for MG are sufficiently effective that the outlook for most people with MG is bright. However, some patients do not find current treatments to be effective and these people may face severe difficulties in managing even basic "activities of daily living" such as brushing one's hair to name just one such activity.

Although current treatments will not cure MG, and there is no treatment that is universally effective or tolerated by all people with MG, most people with MG can expect to have significant improvement in their muscle weakness. In some cases, MG may go into remission for a time, during which no treatment is necessary. There is much that can be done, but still much to be understood. New drugs with greater efficacy and fewer side effects are needed. Research plays an important role in finding new answers and treatments for MG-and the future of MG research has never been brighter.