Muscular Dystrophy Canada
let's make muscles move

Myasthenia Gravis (MG)

What is myasthenia gravis?

Myasthenia gravis is a chronic neuromuscular disorder that produces weakness and abnormally rapid fatigue of the voluntary muscles. MG is considered to be an autoimmune disease because in this disorder the immune system attacks the body. The acetylcholine (ACH) receptor sites at the neuromuscular junction (the point where the nerve endings join the muscle surface) are the targets of this immune reaction.

Who can get myasthenia gravis?

The disorder can affect people of all ages, although it is more common in women between 20 and 30 years of age and in men who are between 40 and 60. It occurs almost twice as often in women.

What causes myasthenia gravis?

The cause of this autoimmune disorder is currently unknown, though MG is not considered to be a genetic disorder.

Normally, a chemical neurotransmitter known as acetylcholine is stored in small vesicles at the end of each nerve. It is released by these vesicles whenever nerve impulses reach the nerve ending. It then forms a chemical bridge to the acetylcholine receptor sites on the muscle side of the neuromuscular junction. As the acetylcholine reaches the receptor sites, a chain of chemical reactions occurs resulting in voluntary muscle contraction.

In MG, the body's immune system has produced antibodies that attack these receptor sites, reducing their number and thus decreasing the ability of the muscle to contract effectively. The result is the weakness and fatigue that is characteristic of myasthenia gravis. The disorder is seldom fatal, though it can be life threatening in situations where muscle weakness interferes with respiration.

What is the role of the thymus gland in myasthenia gravis?

The thymus gland is a small gland that normally produces hormones that affect neuromuscular transmission. Also, the cells of the thymus gland form a part of the body's normal immune system.

Located in the chest beneath the breastbone, the thymus is normally large in infancy and reaches maximum growth by puberty. At that time, it starts to shrink and by late adulthood, it is hardly functional.

In an adult with myasthenia gravis, however, the thymus gland is often abnormally enlarged. The majority of people with MG have some form of abnormality in this gland. Tumours known as thymomas occur in 10 to 15% of these people. They are usually benign, but in some instances may become invasive.

Although the relation of the thymus gland to MG is not totally understood, it appears that the thymus is linked to the acetylcholine receptor antibodies or other substances that interfere with neuromuscular transmission.

How is myasthenia gravis diagnosed?

MG is diagnosed by a neurologist or a specialist in internal medicine. The physician takes a history of the person's symptoms and does a complete physical examination. The diagnosis can be verified by three tests, including an electrical study (EMG), pharmacological testing (Tensilon test) and a blood test to confirm the presence of acetylcholine receptor antibodies.

Is there any cure or treatment?

Presently, there is no cure for myasthenia gravis. However, the use of drugs, surgery on the thymus gland and a treatment known as plasmapheresis, either alone or in combination, have been quite successful in the management of myasthenia gravis. They allow many patients with MG to control their symptoms.

MEDICATIONS

There are several medications used in the treatment of this disorder. These medications are prescribed by a physician. It is important that they are taken as directed and under the ongoing care of the doctor involved. All medical professionals involved in the care of clients with MG should be aware of all medications that are being prescribed.

Anti-cholinesterase or cholinesterase inhibitors are drugs that improve the ACH receptor's ability to retain ACH, thus allowing the muscles to work better. Although they improve weakness, they do not improve the underlying disease. An example of an anti-cholinesterase drug used in the treatment of myasthenia gravis is: Pyridostigmine (Mestinon ™).

Immunosuppressive drugs, which are sometimes known as steroids, are also used. These medications produce marked improvement or remission in many people with MG. They work to suppress the immune response. Two immunosuppressive agents commonly used are: Azathioprine (Imuran™), and Prednisone.

What are the symptoms of myasthenia gravis?

The onset of MG may be sudden, with severe and generalized muscle weakness, but more often the early symptoms are subtle and variable. This may make myasthenia gravis a difficult disorder to diagnose correctly.

Symptoms result from weakness of the voluntary muscles innervated by the ten cranial nerves originating in the brainstem.

Frequently, the first noticeable symptom is weakness of the eye muscles, resulting in droopy eyelids (ptosis) or double vision (diplopia). Other symptoms are related to weakness of the muscles involved in swallowing, chewing, talking or moving the limbs. Symptoms can also include severe fatigue, an unstable or waddling gait, weakness in the arms resulting in an inability to raise the arms over the head, weakness in the hands and fingers and difficulty in swallowing and breathing.

Muscle weakness may develop over a few days or weeks, or remain at the same level for long periods. The severity of the weakness and the degree of muscle involvement varies from person to person, and even in the same person at different times of the day. Weakness tends to worsen with exercise and at the end of the day. It is usually partially alleviated by rest. Symptoms are often more severe during menses and pregnancy.

SURGICAL APPROACH
Thymectomy: Surgical removal of the thymus gland has been shown to favourably affect the course of MG in many people and sometimes leads to remission.

TREATMENT
Plasma Exchange (Plasmapheresis): Plasmapheresis is performed in clients with MG to remove antibodies from the plasma. This may help to reduce symptoms and improve muscle strength. It is used as a short-term intervention in people whose symptoms have worsened suddenly. It may also be used before a thymectomy when a person needs to be in optimum health or with people who have not responded to other therapies. The plasma is exchanged with other fluids that easily combine with the blood and is then returned to the body.

What about research?

Although scientists have learned much about MG so far, researchers are currently trying to determine exactly why the body produces this antibody against the muscle ACH receptor sites. This information will go a long way toward finding a cure for this disorder.

MG can be treated, often very effectively, with immunosuppressive drugs. The disadvantage of these drugs is that they often have unwanted side effects. Researchers are looking at ways to eliminate unpleasant side effects associated with this valuable treatment.

Other researchers are looking for pharmacological treatments that would have fewer side effects than the ones currently in use.

How can I help?

Muscular Dystrophy Canada conducts year-round fund raising campaigns to support our diverse programs. Your gift will help the Association provide the dollars necessary to assist individuals living with neuromuscular disorders, and fund much needed medical research and educational information. Please make a gift through our National office or any Regional or Community Muscular Dystrophy Canada office.

All Muscular Dystrophy Canada Information Sheets are available on our website: www.muscle.ca

Ce feuillet d'information est aussi disponible en français.