

Myasthenia Gravis

Myasthenia Gravis (MG) is a Chronic Autoimmune Neuromuscular condition that causes weakness in certain muscles of the body. MG is caused by an error in the transmission of nerve impulses to the muscles. In MG the immune system makes antibodies that mistakenly attack the connections between nerves and muscles preventing muscles from contracting and resulting in weakness.

What does autoimmune mean?

The immune system (the body's infection-fighting technique) normally makes proteins called "antibodies," which help to prevent or fight infection.

Antibodies are proteins made by the body's immune system when it detects harmful substances. Antibodies may be produced when the immune system mistakenly considers healthy tissue as harmful substance.

What is the neuromuscular junction?

The Neuromuscular junction is the meeting point of a nerve cell and the muscle it controls. Communication happens between the nerve and muscle fibers through the nerve cells called neurons. Due to this communication or transmission of signal, the muscle is able to contract or relax. When the muscles contract, Acetylcholine gets released by the nerve cells. Acetylcholine acts as a chemical messenger that causes our skeletal muscles to contract. The diagram above right shows the neuromuscular junction.

What causes Myasthenia Gravis?

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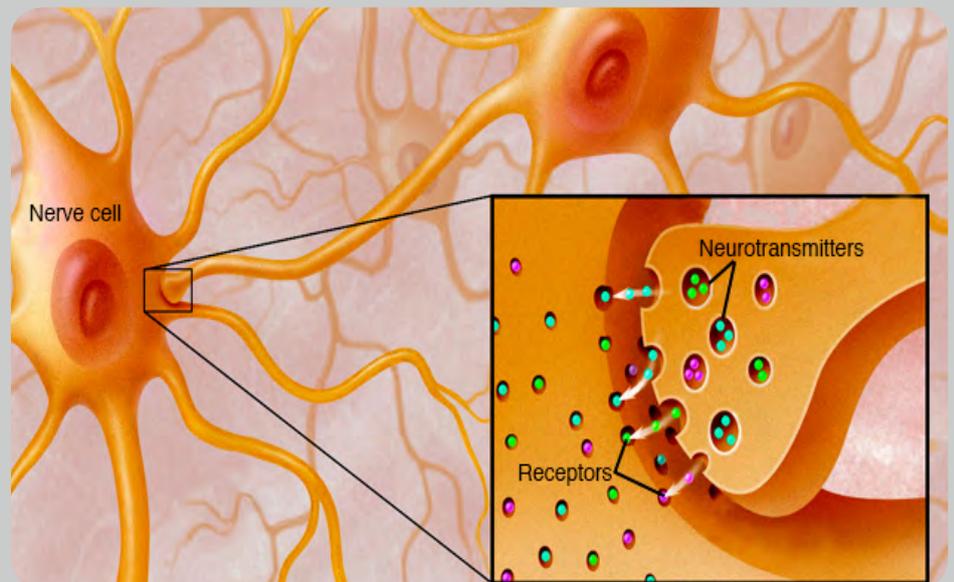


Figure 1: The neuromuscular junction

and resulting in weakness. The diagram on the next page shows the antibodies attacking the junction between nerves and muscle.

Some people may have Myasthenia Gravis that is not caused by antibodies blocking Acetylcholine or the muscle-specific receptor tyrosine kinase. This type of condition is called antibody-negative Myasthenia Gravis.

The role of thymus gland in myasthenia gravis

The thymus gland, lies in the chest area beneath the breastbone (sternum), and plays an important role in the development of the immune system in early life. Its cells form part of the body's normal immune system. The gland

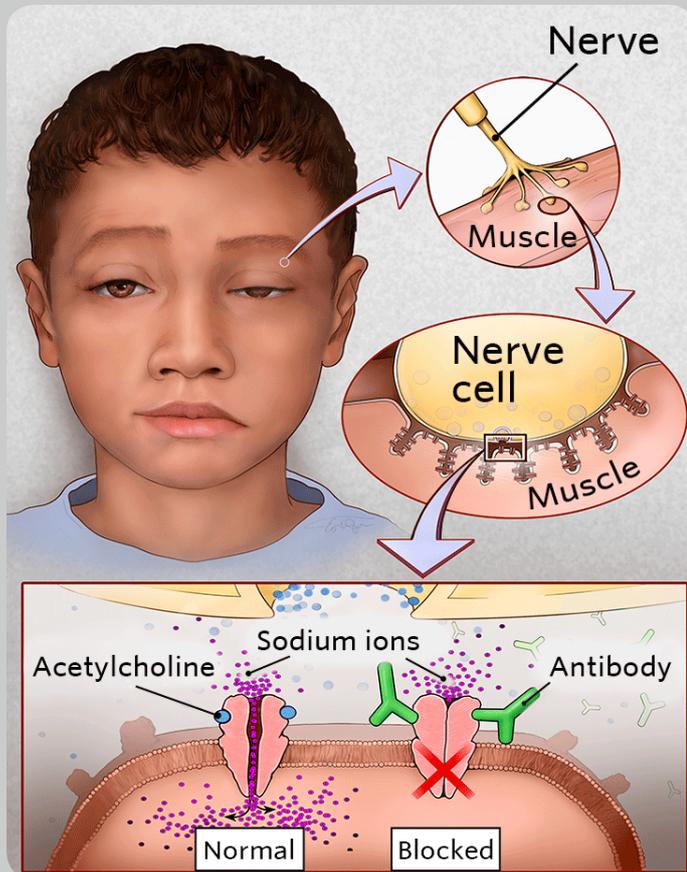


Figure 2: Antibodies attacking the junction between nerves and muscle.

is somewhat larger in infants, grows gradually until puberty, and then gets smaller and is replaced by fat with age. In adults with Myasthenia Gravis, the thymus gland remains large and is abnormal. Some individuals with Myasthenia Gravis develop thymomas (tumors of the thymus gland).

The relationship between the thymus gland and Myasthenia Gravis is not yet fully understood. Scientists believe that the thymus gland may give incorrect instructions to developing immune cells, ultimately resulting in autoimmunity.

What happens if I have Myasthenia Gravis?

The hallmark of Myasthenia Gravis is muscle weakness that increases during periods of activity and improves after periods of rest. Certain muscles such as those that control eye and eyelid movement, facial expression, chewing, talking, and swallowing are often, but not always, involved in the disorder. The muscles that control breathing and neck and limb movements may also be affected.

These include:

- Muscles in the eyelids and around the eyes – If MG only affects these muscles, doctors call it “ocular myasthenia gravis.” About half of all people with MG have this type.

- Jaw muscles
- Arm or leg muscles
- Muscles that help with breathing

What are the symptoms of Myasthenia Gravis?

The main symptom is muscle weakness. It can come and go, and is often worse later in the day. This can result in:

- Ptosis (Droopy eyelids).
- Diplopia (Blurry vision or double vision).
- Trouble chewing food - The jaw muscles might feel tired about halfway through a meal.
- Trouble swallowing.
- Dysarthria (Trouble talking) – A person might speak in a lower voice than usual, or sound like he or she has a cold or stuffy nose.
- Loss of expression on the face.
- Head that feels heavy or drops forward.
- Trouble breathing – A person might feel short of breath, take extra breaths, or feel like it takes a lot of effort to breathe.
- Weakness – It might be hard to lift the arms or legs, open the fingers, or lift a foot.

Who gets Myasthenia Gravis?

Myasthenia Gravis occurs in all ethnic groups and both genders. It most commonly affects young adult women (under 40) and older men (over 60), but can also occur at any age. The condition is not directly inherited nor is it contagious.

What is covered under PMB level of care?

PMBs refer to the benefits as stated in Section 29 (1) (o) of the Medical Schemes Act, No. 131 of 1998 (the Act) Myasthenia Gravis is a PMB condition under Diagnosis and Treatment Pair (DTP) code 513A.

This DTP refers to Myasthenia gravis as muscular dystrophy; neuro-myopathies NOS. The treatment component specified for this DTP according to the PMB Regulations is Initial diagnosis; initiation of medical management; therapy for acute complications and exacerbations. The following should therefore be paid according to the PMB Regulations:

Diagnosis

The doctor will record the patient’s medical history, do a physical and detailed neurological examination to understand the symptoms. If the doctor suspects Myasthenia Gravis, several tests can be performed to confirm the diagnosis. Not all patients qualify for all the tests mentioned below. The investigations are done subject to the discretion of the treating doctor.

The tests can include:

- Blood tests for certain antibodies that are found in people with MG such as acetylcholine receptor antibodies.
- Electrical tests of nerves and muscles – Repetitive nerve stimulation. This is a nerve conduction study in which doctors attach electrodes to your skin over the muscles to be tested. Doctors send small pulses of electricity through the electrodes to measure the nerve's ability to send a signal to your muscle. To diagnose Myasthenia Gravis, doctors will test the nerve many times to see if its ability to send signals worsens with fatigue.
- Imaging tests, such as Computerized Tomography (CT) or Magnetic Resonance Imaging (MRI) scans – Imaging tests can show changes, including a tumor on the thymus gland.
- Ice pack test – During this test, a doctor puts a cool pack on the eyelids. If the eyelids open better after resting under the cool pack, this could be a sign of MG.
- Pulmonary function testing - which measures breathing strength, helps to predict whether respiration may fail and lead to a Myasthenia crisis.

How is Myasthenia Gravis treated?

Initiation of treatments, which may include:

- Medicines to treat muscle weakness, such as pyridostigmine.
- Medicines that treat the immune system over time, such as prednisone or azathioprine. These medications improve muscle strength by suppressing the production of abnormal antibodies.
- Fast-acting immune system treatments, such as:
 - A medicine called “intravenous immune globulin” (IVIG) – high-dose intravenous immune globulin, which temporarily modifies the immune system by infusing antibodies from donated blood.
 - Plasma exchange (also called “plasmapheresis”) – a procedure in which serum containing the abnormal antibodies is removed from the blood while cells are replaced.
- Surgery to remove the thymus gland in patients with thymoma (tumor of the thymus gland / abnormal tissue of the thymus gland) or hyperplasia of the thymus gland.
- Treatment options also include methotrexate, mycophenolate mofetil and cyclosporine.

As the PMB Regulations stipulate initiation of treatment, the chronic use of the abovementioned treatments is not included and the medical scheme does not have to fund the chronic medicine in full.

Therapy for exacerbations and acute complications such as pneumonia and respiratory failure must however be funded as PMB level of care by the medical scheme.

These therapies may be used to help individuals during especially difficult periods of weakness. A Neurologist will determine which treatment option is best for each individual depending on the severity of the weakness, the kind of muscles are affected, as well as the individual's age and other associated medical problems.

If MG attacks the muscles that help with breathing, it can cause severe breathing problems. These patients are usually treated in the Intensive Care Unit (also called the “ICU”) and may require assisted ventilation.

Children with MG can take some of the same medicines prescribed for adults, at age appropriate doses. However, some medicines used to treat MG can cause more serious problems in children if used for a long time. Surgery to remove the thymus gland is safe for children and often works well.

What is a Myasthenic crisis?

A Myasthenic Crisis occurs when the muscles that control breathing weaken to the point that ventilation is inadequate, creating a medical emergency and requiring a respirator for assisted breathing. Respiratory failure is included in the PMBs and the medical scheme must fund the medical treatment, oxygen and ventilation in full.

What else should I know about Myasthenia Gravis?

People who have MG that affects more than just the eyes can have serious problems if they get the flu or pneumonia. For this reason, it is especially important that they get the flu vaccination every year and the pneumonia vaccine as per the national vaccination guideline.

Some medicines can make MG worse. Talk to your doctor or nurse before you take any medicines, including over-the-counter medicines. If you get a prescription for a new medicine, ask if it is safe to take when you have MG.

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WHAT ARE PRESCRIBED MINIMUM BENEFITS?

Prescribed Minimum Benefits (PMBs) are defined by law. They are the minimum level of diagnosis, treatment, and care that your medical scheme must cover – and it must pay for your PMB condition/s from its risk pool and in full. There are medical interventions available over and above those prescribed for PMB conditions but your scheme may choose not to pay for them. A designated service provider (DSP) is a healthcare provider (e.g. doctor, pharmacist, hospital) that is your medical scheme's first choice when you need treatment or care for a PMB condition. You can use a non-DSP voluntarily or involuntarily but be aware that when you choose to use a non-DSP, you may have to pay a portion of the bill as a co-payment. PMBs include 270 serious health conditions, any emergency condition, and 25 chronic diseases; they can be found on our [website](#)

The Communications Unit would like to thank the Clinical Unit for assisting with this edition of CMScript

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