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# MYASTHENIA GRAVIS LEXICON

LEXICON



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# Introduction

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In collaboration with clinical, patient, and industry stakeholders, the MGFA has developed the Myasthenia Gravis (MG) Lexicon, a comprehensive educational resource designed to standardize terminology and promote effective communication across the MG community. Through its collection of key MG terms and concepts, the lexicon serves as the foundation for a shared, unified language that brings clarity, establishes consistency, and enhances mutual understanding.

## Our Mission

The MGFA's purpose is to enhance the lives of those affected by MG by creating connections, improving care, and supporting MG research. The MG Lexicon supports this mission by creating a shared language that strengthens dialogue between patients, caregivers, clinicians, researchers, and industry partners.

## Lexicon

The MG Lexicon is a comprehensive glossary of MG terminology, curated in collaboration with clinicians, patient advocates, and industry partners. It is designed to help community members “talk the same language” when discussing, diagnosing, and understanding MG. By providing clear definitions and context, the Lexicon ensures consistent, accessible communication for all stakeholders.

## Key Audience

The Lexicon is intended for a broad audience, including patients, caregivers, healthcare professionals, researchers, and industry collaborators. Its purpose is to simplify complex MG terminology, supporting both clinical discussions and community education.

## Scope and Focus

The Lexicon focuses on key MG terminology, treatment options, and disease concepts, acknowledging that treatments and approaches continue to evolve.

## Lexicon Development Process

The creation of the MG Lexicon was driven by the MGFA Stakeholder Roundtable meetings, which brought together experts across the MG community. Input from industry partners, clinicians, and patient representatives shaped the content, ensuring that the Lexicon reflects a balanced, informed, and practical approach to MG terminology. MGFA is thankful for the participation and partnership of all those involved in creating this resource.

## How to Use this Resource

Each term, phrase, or medical term on pages 3 through 6 are actually live links to definitions. Each term includes a small arrow. Click or tap on each word or phrase to obtain a definition and more detailed information. The actual definitions for MG-related words and phrases can be found on pages 7 through 18 in the Lexicon.



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# Diagnosing MG

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A healthcare provider who suspects myasthenia gravis (MG) will assess certain symptoms and conduct a thorough neurological examination, followed by blood and/or electrodiagnostic testing, to help confirm or eliminate a diagnosis. Below are the symptoms an individual with MG may have, as well as the various tests that can be used to help determine a proper diagnosis.

## MG Symptoms

**Bulbar weakness ▶**

**Diplopia ▶**

**Dysarthria ▶**

**Dysphagia ▶**

**Dysphonia ▶**

**Dyspnea ▶**

**Fatigable muscle weakness ▶**

**Fluctuating muscle weakness ▶**

**Ptosis ▶**

*\*Other symptoms of myasthenia gravis include changes in facial expression and weakness in the neck and limb muscles. Furthermore, MG presents differently from patient to patient, which is why many in the community call MG a "snowflake disease." Even though individuals share the same diagnosis, the severity and range of symptoms differ.*

## Tests Used to Diagnose MG

### PHYSICAL TESTING

**Dynamometry ▶**

**Ice pack test ▶**

**Neurological exam ▶**

### BLOOD ANALYSIS

**Antibody serology tests ▶**

### ELECTRODIAGNOSTIC TESTING

**Repetitive Nerve Stimulation (RNS) ▶**

**Single-Fiber Electromyography (EMG) ▶**

### IMAGING

**Computed Tomography Scan (CT Scan) ▶**

**Magnetic Resonance Imaging (MRI) ▶**

# Newly Diagnosed

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Once a diagnosis is confirmed by a healthcare professional, there will be general, basic terms that an individual with myasthenia gravis will become familiar with as they embark on their MG journey. Getting acquainted with an MG diagnosis can take time, but learning the types of MG, the different antibodies associated with MG, and how the disease impacts the body on a basic level can aid in one's understanding of the disease. Below are the various types and subtypes of MG, the specific antibodies related to myasthenia gravis, and parts of the body involved in the MG disease process.

## Physiology Terms

- Autoantibody ▶
- Atrophy ▶
- Autoimmune disease ▶
- B Cells ▶
- Complement system ▶
- Cytokines ▶
- Immunoglobulin ▶
- Immune system ▶
- MGFA clinical classification ▶
- Neonatal FC receptors (FcRn) ▶
- Neuromuscular disorder ▶
- Neuromuscular junction (NMJ) ▶
- Neurotransmitter ▶
- Receptor sites ▶
- Skeletal muscle ▶
- T cells ▶
- Tolerance ▶
- Thymic hyperplasia ▶
- Thymoma ▶

## MG Types, Subtypes, and Related Conditions

Myasthenia gravis can be categorized in a variety of ways and can be understood based upon disease onset, specific muscle groups, antibody types, autoimmunity, and more.

### DISEASE ONSET

- Early-onset myasthenia gravis ▶
- Juvenile myasthenia gravis ▶
- Late-onset myasthenia gravis ▶

### MG SEROTYPE

- Acetylcholine receptor antibody negative (AChR-) ▶
- Acetylcholine receptor antibody positive (AChR+) ▶
- Lipoprotein receptor-related protein 4 (LRP4) ▶
- Muscle-specific tyrosine kinase positive (MuSK) ▶
- Seronegative myasthenia gravis\* ▶

*\*Seronegative myasthenia gravis refers to individuals who exhibit myasthenia gravis symptoms but do not have detectable antibodies associated with MG in their blood. Researchers believe that this type of MG still derives from an autoimmune issue, but antibodies involved have simply not been identified yet.*

## SUBTYPES\*

**Generalized myasthenia gravis (gMG) ▶**

**Ocular myasthenia gravis (OMG) ▶**

*\*Ocular myasthenia gravis involves symptoms that only affect the eye muscles. In generalized myasthenia gravis, symptoms can affect muscles throughout the body, including the eye muscles.*

## INHERITED

**Congenital myasthenic syndrome (CMS) ▶**

## OTHER

**Lambert Eaton (myasthenic) syndrome (LEMS) ▶**

**Serious adverse event (SAE) ▶**

**Side effect ▶**

**Specialty pharmacy ▶**

**Transient neonatal myasthenia gravis ▶**

## THYMUS GLAND CHARACTERIZATION

**Non-thymomatous myasthenia gravis ▶**

**Thymomatous myasthenia gravis ▶**

# Living With and Managing MG

Learning to live with MG effectively is an important step to take after an MG diagnosis. With time, individuals living with MG will become acquainted with medical jargon that is either specific to myasthenia gravis or relates to MG care. Below are various terms and concepts one may encounter when managing an MG diagnosis.

## COMMONLY USED MG SCALES

- Activities of Daily Living Scale (ADL) ▶
- MG Impairment Index (MGII) ▶
- MG Quality of Life 15 revised (MG QOL15r) ▶
- Myasthenia Gravis Composite Score (MGC) ▶
- Myasthenia Gravis Post-Intervention Status (PIS) ▶
- Quantitative Myasthenia Gravis Score (QMG) ▶
- Quantitative Myasthenia Gravis Revised (QMGR) ▶

## MEDICAL AND ALLIED HEALTH PROFESSIONALS

- Advance practice provider (APP) ▶
- Emergency medical technicians (EMT) ▶
- Healthcare provider (HCP) ▶
- Neurologist ▶
- Neuromuscular specialist ▶
- Occupational therapist ▶
- Ophthalmologist ▶
- Neuro-ophthalmologist ▶
- Intensivist ▶
- Pediatric neurologist ▶
- Pulmonologist ▶
- Paramedic ▶
- Pathologist ▶
- Pharmacist ▶
- Physical therapist ▶
- Primary care provider ▶
- Respiratory therapist ▶
- Social worker ▶
- Speech language pathologist ▶
- Thoracic surgeon ▶

## MG COMPLICATIONS

- Cholinergic crisis ▶
- Exacerbation ▶
- Myasthenic crisis ▶

## MG TREATMENTS\*

- Antibody ▶
- Biologic ▶
- Biosimilar ▶
- BiPAP ▶
- CAR T-cell ▶
- Cell therapy ▶
- Cholinesterase inhibitors ▶
- Complement inhibitors ▶
- Corticosteroid ▶
- FcRn blockers ▶
- Ig degraders ▶
- Immunomodulators ▶
- Immunosuppression ▶
- Immunosuppressants (ISTs) ▶
- Immunoabsorption ▶
- Intravenous immunoglobulin (IVIG) ▶
- Noninvasive ventilation (NIV) ▶
- Nonsteroidal immunosuppressants (NSISTs) ▶
- Plasmapheresis / Therapeutic plasma exchange (TPE), (PLEX) ▶
- siRNA ▶
- Subcutaneous immunoglobulin ▶
- Thymectomy ▶

\*New treatment paradigms are being studied and trialed. As such, this list is likely to expand. In addition, some of these treatments are still being studied as a treatment for MG.

## OTHER TERMS:

### CLINICAL TRIAL PHASES

- Pre-clinical phase ▶
- Phase I ▶
- Phase II ▶
- Phase III ▶
- Phase IV ▶

# MG Definitions

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## A

**AChR:** Acetylcholine is a chemical that helps muscles contract. It acts as a messenger between nerves and muscles. People who have myasthenia gravis often make an abnormal protein called acetylcholine receptor antibody.

**Acetylcholine (ACh):** A neurotransmitter, or a chemical, that carries messages between the nerve and muscle.

**Acetylcholine Receptor antibody positive (AChR+):** A type of myasthenia gravis that is diagnosed by the presence of acetylcholine receptor (AChR) antibodies. Acetylcholine receptor antibodies cause the breakdown of the receptors, which blocks of acetylcholine's binding to the receptors. Approximately 70-85% of people with MG have AChR+ MG.

**Acetylcholine receptor antibody negative (AChR-):** A term used to identify people with myasthenia gravis who do not have detectable antibodies to the acetylcholine receptor when tested. This group may include patients with antibodies to the MuSK and LRP4 proteins, or those who do not have any detectable antibody by currently used laboratory assays. This term has a different meaning than seronegative myasthenia gravis.

**Advance practice provider (APP):** A medical professional who is not a doctor, but has special training and is licensed to diagnose, treat, and manage many common medical conditions. Advanced practice providers can perform physical exams, order laboratory tests and imaging tests, review test results, prescribe medications, perform or assist with certain procedures (such as biopsies), and make referrals to other health care providers. They may also provide information about screening tests and give counseling on living a healthy lifestyle, such as changes in diet and exercise and quitting smoking. Examples of advanced practice providers are nurse practitioners and physician assistants.

**Antibody:** An antibody is a protein produced by the immune system to help identify and neutralize foreign substances such as bacteria, viruses, and toxins. In autoimmune diseases, the autoimmune antibody, also known as an autoantibody, is a type of protein produced by the immune system that mistakenly targets and attacks the body's own tissues and organs. Synonym: immunoglobulin.

**Antibody serology test:** A test that checks for the presence or level of specific antibodies in the blood.

**Apheresis:** A process that uses a machine to separate blood into different components: plasma (the fluid that carries blood cells) and cells (white blood cells, red blood cells, and platelets). Plasmapheresis is a form of apheresis that is used in myasthenia gravis to remove the plasma containing disease-causing autoantibodies (e.g. AChR antibody).

**Atrophy:** The wasting or decrease in size of an organ, tissue, or body part. There are various types of atrophy, including muscle atrophy, which is when muscle tissue thins, breaks down, and decreases in size.

**Autoantibody:** A type of protein made by the immune system that mistakenly targets the body's healthy tissues and organs instead of bacteria and viruses.

**Autoimmune disease:** A condition in which the body's immune system mistakes its own healthy tissues as foreign and attacks them.

# B

**B cells:** A type of white blood cell that defends the body against disease. Their main function is to produce antibodies. B cells are produced from precursor cells in the bone marrow and go through several stages of maturation to become “plasma” cells, which can produce antibodies. Synonym: B lymphocyte.

**B-Cell depletion therapy:** B-cell depletion is a treatment used for autoimmune diseases. The goal of B-cell depletion is to lower the number of B cells in the body. Because B cells produce antibodies, which can mistakenly target the body's own tissues in autoimmune diseases, depleting the B cells can decrease production of harmful antibodies.

**Biologic therapy:** Biologic therapies are a type of medication made from living organisms, or parts of them, to help fight diseases. These treatments often use proteins, cells, or antibodies made from living cells. Biologic therapy may help a person's own immune system work better to directly target and destroy disease-causing cells. Biologic medications are typically more complex than other medications because of how they are produced. Examples of biologic therapies include vaccines, types of insulin, and monoclonal antibodies.

**Biosimilars:** A biosimilar is a medicine that is designed to be very similar to another biologic medicine that has already been approved. Both biologic therapies and biosimilars are made from living things, such as cells. Biosimilars usually cost less than the original approved medicine, which makes them easier for people to afford. Even though they are not identical, biosimilars are tested extensively to make sure that they are as safe and efficacious as the product to which they are similar.

**BiPAP:** Bilevel positive airway pressure (BiPAP) is a type of noninvasive ventilation that providers might use if a patient can breathe on one's own but is not getting enough oxygen or cannot get rid of carbon dioxide. This is a preferred method over CPAP because it may be more comfortable for patients.

**Bulbar weakness:** A type of weakness within specific facial muscles that may cause difficulty with speaking, chewing, swallowing, and holding up the head. Examples include slurred or nasal speech, choking, and coughing with drinking liquids.

# C

**CAR-T cell therapy:** A type of therapy that takes a patient's T cells (a key part of the immune system that can recognize and destroy invading organisms) and reprograms them to attack a specific target. In myasthenia gravis, the targeted cells are those that make damaging antibodies.

**Cell therapy:** A type of treatment where healthy cells are injected or transplanted into a patient's body. The goal of cell therapy is to help replace or repair damaged cells. One example of cell therapy is CAR-T.

**Central fatigue:** Central fatigue is a feeling of tiredness and lack of energy that is not related to muscle weakness or pain. This type of fatigue can interfere with mental and physical activities, making it hard to think clearly or focus. Central fatigue may also show as feeling unmotivated to do physical activity even when muscles are working well.

**Cholinergic crisis:** A cholinergic crisis is a medical event that occurs when there is a high level of acetylcholine in the body. In MG, some medications used to treat the disease work to increase acetylcholine, but if these medications are taken in high doses, it can lead to too much acetylcholine and potentially results in a cholinergic crisis. This type of crisis is rare with acetylcholinesterase inhibitor (e.g. pyridostigmine) doses under 120mg three times daily (total dose 360mg). Symptoms of a cholinergic crisis may include increase saliva or secretions, diarrhea, muscle cramps or weakness, or blurry vision. These may be hard to distinguish from a myasthenic crisis.

**Cholinesterase inhibitors:** Cholinesterase inhibitors (also called acetylcholinesterase inhibitors) are a class of medications that slow the normal breakdown of the neurotransmitter acetylcholine. Slowing the normal breakdown increases the levels of acetylcholine within the body, resulting in improved muscle strength and better communication between nerves and muscles.

**Clinical trial phases:** Clinical trials are research studies used to determine if medications are both safe and effective. Before clinical trials begin in humans, preclinical studies are often conducted in animals to evaluate the biological activity and safety of the therapy. Clinical trials have different phases to study varying aspects of the medication. In rare diseases such as MG, the number of patients enrolled in each of these study phases may be smaller than in studies for other diseases.

- **Pre-clinical phase:** The stage before testing a new drug or treatment on people. During this time, researchers do experiments in labs and often use animals to learn about the treatment's safety and effectiveness.
- **Phase I:** This phase involves researchers testing a drug or treatment in a small group of healthy people for the first time. The study determines how a drug is absorbed, distributed, metabolized, and excreted as well as the duration of its action.
- **Phase II:** In this phase, controlled trials of a larger number of patient volunteers (people with the disease) assess a drug's effectiveness and safety.
- **Phase III:** This phase usually involves a larger group of patients in clinics and hospitals. Physicians monitor patients closely to confirm efficacy, identify adverse events, and compare the new drug to standard or similar treatments.
- **Phase IV:** After a treatment has been approved and is being marketed, it is studied in a Phase IV clinical trial. Researchers track safety and evaluate side effects of the new treatment as it is used in the general population. Thousands of people are involved in a Phase IV trial.

**Complement system:** The complement system is part of the immune system, made up of a group of proteins that work to keep the body healthy. The complement system works to target and remove invaders like viruses and bacteria and activates inflammation in the body to prevent infection. In autoimmune diseases, the complement system can become overly active and may not only destroy foreign invaders but also normal, healthy tissue (including the neuromuscular junction).

**Complement inhibitors:** Complement inhibitors are a class of medications that are used in the treatment of MG. This class is a type of immunotherapy used to treat many inflammatory conditions. They work by stopping the activity of the complement system, which is a part of the body's immune system. As a class, these medications have a black box warning from the FDA because they require meningococcal vaccination before starting treatment as part of a Risk Evaluation and Mitigation Strategy (REMS) program.

**Complete stable remission:** A state in which a patient shows no signs or symptoms of disease activity over an extended period.

**Congenital myasthenic syndromes (CMS):** A group of rare disorders that all involve disruption at the neuromuscular junction. These disorders are caused by genetic changes affecting the neuromuscular junction. Unlike autoimmune-driven myasthenia gravis, CMS does not involve the immune system attacking the body.

**Corticosteroid:** Often referred to as just "steroids," corticosteroids are a class of medication that is used to reduce the abnormal immune response in myasthenia gravis. Corticosteroids can help manage symptoms by decreasing inflammation and immune activity.

**Crisis:** See **myasthenic crisis**.

**CT scan:** Computed Tomography Scan (CT scan), is a medical imaging procedure that uses X-rays and computer processing to create detailed, cross-sectional images of inside the body. It is also known as a CAT scan (Computed Axial Tomography).

**Cytokines:** Cytokines are small proteins that help cells communicate with each other in the body. They play a key role in the immune system, helping to control how the body responds to infections, inflammation, and other signals. Cytokines act like messengers that inform cells what to do.

# D

**Diplopia:** Double vision.

**Dysarthria:** Slurred speech.

**Dysphagia:** Difficulty swallowing.

**Dysphonia:** A general term used to describe changes in voice quality or production. Examples may include a nasal-sounding voice or low voice volume.

**Dyspnea:** The feeling of uncomfortable breathing or shortness of breath that can vary in intensity.

**Dynamometry:** A method used to measure muscular strength, power, and/or endurance. This assessment is often done by using a hand-held device and is usually performed by a knowledgeable physical therapist.

# E

**Early-onset myasthenia gravis:** A type of myasthenia gravis where the first symptoms begin before the age of 50.

**Electrodiagnostic testing:** A type of testing used to identify or diagnose injuries or diseases that affect the peripheral nervous system and skeletal muscles. Examples of EMG testing include nerve conduction studies (NCS) and needle electromyography (EMG), which help to evaluate how well nerves and muscles function.

**Emergency medical technicians (EMT):** An emergency medical technician (EMT) is a medical professional who gives emergency care to people outside of or on the way to the hospital. They can give basic medical and first-aid care until a person can get help at a hospital or other medical facility.

**Exacerbation:** A temporary worsening of myasthenia gravis symptoms. Exacerbations may be triggered by various factors, such as infection, reduction of MG medications, the use of high-dose corticosteroids, or a medication used for other purposes that may have increased MG symptoms. In the MG community, the word “flare” is a term that individuals sometimes use to describe the symptoms of an exacerbation.

# F

**FcRn blockers/inhibitors:** FcRn blockers, a class of medications used in MG treatment, are also known as FcRn inhibitors. These medications are monoclonal antibodies that block the function of the neonatal Fc receptor, which is normally responsible for recycling IgG (a type of antibody). By stopping recycling, FcRn blockers lower the levels of circulating antibodies.

**Fatigable muscle weakness:** Weakness of muscles that can vary in severity and usually worsens with physical activity and improves with rest. Fatigable muscle weakness can happen with bulbar symptoms in the face or other muscle groups, like the arms and legs.

**Fluctuating weakness or symptoms:** Fluctuating weakness is like fatigable weakness but refers to the variability experienced throughout time. In MG patients, weakness tends to change throughout the day or worsen as the day progresses. These changes may happen from hour to hour, day to day, week to week, or beyond. Fluctuations can be applied to other MG symptoms in addition to muscle weakness.

# G

**Generalized myasthenia gravis (gMG):** A form of myasthenia gravis in which weakness affects bulbar muscles (muscles that control speech, swallowing, and chewing), respiratory (breathing) muscles, neck, and limb muscles to varying degrees of weakness. Sometimes people with generalized myasthenia gravis have ocular muscle weakness as well. Seventy-five percent of patients with MG will have gMG.

# H

**Healthcare provider (HCP):** A healthcare provider is a person or entity that provides medical care or treatment. Healthcare providers include doctors, nurse practitioners, physician assistants, pharmacists, midwives, radiologists, labs, hospitals, urgent care clinics, medical supply companies, and other professionals, facilities, and businesses that provide such services.

# I

**Ice Pack Test:** A test used to diagnose MG if a patient has a droopy eyelid. An ice pack is placed on the drooping eyelid for two minutes. If the cooling of the ice pack temporarily improves the drooping eyelid, there is likely support for a diagnosis of MG. This is because the cool temperature may improve neuromuscular transmission in the state of MG.

**Ig degraders:** An Ig (or immunoglobulin) degrader is a molecule designed to target and break down immunoglobulins, which are antibodies produced by the immune system. These degraders can be used to reduce the levels of specific antibodies in the body, which can be beneficial in treating certain autoimmune diseases and other conditions where pathogenic antibodies play a role.

**Immune checkpoint inhibitor-induced myasthenia gravis:** A type of myasthenia gravis that can occur as a side effect of treatments called immune checkpoint inhibitors. These medications are designed to help the immune system attack cancer cells, but in some cases, they can trigger symptoms of myasthenia gravis in patients.

**Immune system:** A complex system of cells, tissues, and organs that work together to defend the body against infection and disease. The immune system can help to identify and target harmful invaders, like bacteria and viruses, to keep people healthy.

**Immunizations:** Immunization is the process of becoming protected against a disease with a vaccine, also called vaccination. Vaccines help to stimulate the immune system to recognize and fight specific diseases.

**Immunoadsorption:** Immunoadsorption is a procedure similar to plasma exchange that removes harmful (pathogenic) antibodies from the bloodstream.

**Immunoglobulin (IVIg):** See **antibody**.

**Immunomodulators:** Medications that change how the immune system works, helping it to function more effectively.

**Immunosuppressants (ISTs):** Drugs that suppress the immune response of an individual. They are often used to prevent the immune system from attacking the body's own tissues in autoimmune diseases.

**Immunosuppression:** A state in which the immune system is suppressed or weakened, which lowers the body's ability to respond to foreign invaders and makes a person more vulnerable to illness or infections.

**Intravenous Immunoglobulin (IVIG):** A treatment that uses purified immunoglobulin from healthy donors. This treatment is given through a patient's vein. The exact mechanism of how IVIG works in successfully treating myasthenia gravis is not entirely understood, however; it seems to affect the function or the production of antibodies in the immune system.

**Intensivist:** A physician who specializes in the care and treatment of patients in an intensive care setting.

## J

**Juvenile myasthenia gravis:** A subtype of early onset myasthenia gravis that begins in children younger than 18 years of age. About half of juvenile cases have an onset before the age of 15. In some places, juvenile MG may be used interchangeably with adolescent MG, but the two have different meanings in the context of clinical trials.

## L

**Lambert Eaton (myasthenic) syndrome:** Lambert Eaton syndrome (LES) – or Lambert Eaton myasthenic syndrome (LEMS) – is an autoimmune condition with features similar to MG that is due to impaired neuromuscular transmission. LEMS often results from antibodies that target the presynaptic neuromuscular junction, while MG targets the postsynaptic muscle membrane. Both LEMS and MG can cause fatigable muscle weakness, often starting in the limbs. Unlike MG, it is less common in LEMS to see issues with eye movements and bulbar movement. LEMS may be associated with cancer in about 60% of patients.

**Late-onset myasthenia gravis:** Myasthenia gravis with onset of first symptoms after 50 years of age.

**Lipoprotein receptor-related protein 4 antibody-positive (LRP4+):** A type of myasthenia gravis that is diagnosed by the presence of LRP4 antibodies. LRP4 is a protein important for maintenance of the neuromuscular junction. Approximately 1-5% of people with MG have LRP4+ MG.

## M

**Mechanism of Action (MOA):** Mechanism of action describes how a specific drug works in the body to produce its effects. It describes the specific ways the drug interacts with cells, tissues, or systems to help treat a condition. Understanding the mechanism of action can help patients and healthcare professionals understand how a medication may work and what to expect from use.

**MGFA clinical classification:** The MGFA Clinical Classification is a tool used, often in clinical research and clinical practice, to understand the severity of a patient's disease based on clinical features. The classes are divided into five different groups. Some groups have subclasses of A and B, where A means the limb and trunk muscles are more involved than bulbar muscles, and B means the bulbar muscles are more involved than limb and trunk. A higher class of MGFA clinical classification indicates more severe MG.

- **Class 0:** Clinical remission.
- **Class I:** Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal. Class II: Mild weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IIA.** Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IIB.** Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
- **Class III:** Moderate weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IIIA.** Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IIIB.** Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
- **Class IV:** Severe weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.
  - **IVA.** Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.
  - **IVB.** Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
- **Class V:** Defined as intubation, with or without mechanical ventilation, except when employed during routine postoperative management. The use of a feeding tube without intubation places the patient in class IVB.

**MG Impairment Index (MGII):** The MG Impairment Index is a tool to assess the severity of MG through patient self-reporting (22 items) and clinical examination (6 items). It assesses impairments in various areas, including vision (ocular), bulbar, breathing (respiratory), and movement. Higher scores in the MGII indicate more severe impairments.

**MG Quality of Life Scale Revised (MG-QOL-15R):** The MG-QOL-15R is a questionnaire of 15 items that patients complete to assess how MG impacts their physical, psychological, and social well-being. The revised version reduced the number of responses for each item from 4 to 3. Each item is scored 0 to 2 for a maximum score of 30 points. Higher scores indicate more severe impairment in quality of life.

**Minimal symptom expression (MSE):** When a patient has an MG-ADL total score of 0-1 or MG-QOL-15R total score of 0-3. This is a term used to identify patients with measured scores that experience minimal symptoms.

**Monoclonal antibody:** A type of antibody made in a lab used to treat various medical conditions. Monoclonal antibodies are designed to find and destroy harmful materials in the body, like germs or specific cells. Healthcare providers use monoclonal antibodies to help treat diseases by helping the immune system fight infections or by targeting and destroying harmful cells.

**MRI:** Magnetic Resonance Imaging (MRI) is a noninvasive medical imaging procedure that uses a powerful magnetic field and a computer to produce detailed images of the organs and tissues inside the body.

**Muscle-specific tyrosine kinase antibody positive (MuSK+):** A type of myasthenia gravis that is diagnosed by the presence of MuSK antibodies. MuSK is an important protein for the maintenance of the neuromuscular junction. Approximately 1-10% of people with MG have MuSK+ MG.

**Myasthenic crisis:** A severe and life-threatening exacerbation of MG in which the muscles responsible for breathing (respiratory muscles) fail. A myasthenic crisis often requires respiratory support, such as noninvasive positive pressure ventilation or mechanical ventilation (intubation). Some patients may also experience severe bulbar weakness, which impacts the muscles involved in swallowing. When this occurs, the airway may be obstructed, and intubation or mechanical ventilation may be needed.

**Myasthenia Gravis Activities of Daily Living Scale (MG-ADL):** The MG-ADL is an eight-item questionnaire that patients complete to assess the severity of their MG symptoms and impact on activities of daily living. All items ask about the experience of patients over the previous seven days. The MG-ADL is a patient reported outcome tool that provides a score, which helps providers to understand the severity of MG symptoms. MG-ADL scores range from 0-24, with higher scores indicating more severe symptoms and impact.

**Myasthenia Gravis Composite Score (MGC):** The MG Composite Score indicates disease severity based on impairments of body function and structure. This 13-question measure is derived from components of the MG-ADL score, the Quantitative Myasthenia Gravis score and the MG-Manual Muscle Testing Score. The score range is 0-50, with higher scores indicating higher severity.

## N

**Neonatal Fc receptor (FcRn):** The neonatal Fc receptor (FcRn) is a protein that helps distribute, transport, and maintain IgG (immunoglobulin) in the body.

**Neurological exam:** A physical examination performed by a medical professional to better understand the function of a patient's nervous system. A neurological exam can include various tests to check mental status, speech, strength, coordination, muscle tone, reflexes, and more.

**Neurologist:** A doctor who specializes in diagnosing and treating disorders that affect the brain, spinal cord, and peripheral nervous system.

**Neuromuscular disorder:** A condition that impacts the functioning of nerves, muscles, or the connection between nerves and muscles (also see neuromuscular junction). These disorders may lead to weakness or loss of control over muscle movements.

**Neuromuscular junction (NMJ):** A specialized area where nerve endings come very close muscle fibers. This is where signals are sent from the nerve to the muscle. When a nerve impulse travels down the nerve to the neuromuscular junction, it releases a chemical called a neurotransmitter, which sends a signal to the muscle to contract. In myasthenia gravis, antibodies disrupt this signaling process, resulting in muscle weakness.

**Neuromuscular specialist:** Neuromuscular specialists are healthcare providers that evaluate and diagnose disorders affecting motor neurons, nerves, neuromuscular junctions, and muscles. They are experts in conditions that impact muscle control and movement.

**Neuro-ophthalmologist:** A physician who specializes in vision or eye problems related to conditions of the brain and nervous system. They diagnose and treat issues that involve both the eyes and neurological functions.

**Neurotransmitter:** A chemical “messenger” in the brain that helps carry signals between nerves and other cells. These chemical messengers are released by the end of a nerve (nerve terminal) and travel across a junction (known as a synapse) to reach another nerve ending or muscle fiber. Acetylcholine is an example of a neurotransmitter that is used at the neuromuscular junction and is one of several different neurotransmitters in the body that facilitate communication between nerves and muscles.

**Noninvasive ventilation (NIV):** A way to provide breathing support through a mask or nasal prongs, instead using an invasive artificial airway like an endotracheal tube (which is inserted through the mouth into the windpipe, or “trachea”) or tracheostomy (a tube inserted from the front of the neck directly into the windpipe). Noninvasive ventilation is often used to help people breathe more easily without invasive options.

**Non-thymomatous myasthenia gravis:** A type of myasthenia gravis in which there is no thymoma (tumor of the thymus gland) present.

**Nonsteroidal immunosuppressants (NSISTs):** A class of medications used to suppress the immune system without the use of corticosteroids.

## O

**Occupational therapist:** A healthcare professional trained to help individuals improve their ability to perform daily tasks, such as getting dressed or brushing one’s teeth.

**Ocular myasthenia gravis (oMG):** A type of myasthenia gravis in which weakness is limited to the eye muscles, including the eyelid and the muscles that control eye movement. Common symptoms of ocular MG include diplopia and ptosis. Approximately 20% of patients with MG will have ocular MG.

**Off-label:** A medication is considered “off-label” when a healthcare provider prescribes a drug to be used for a condition that is different from what was approved by the FDA. This practice is common and legal within the United States.

**On-label:** A medication is considered “on-label” when a healthcare provider prescribes a drug to be used in the same condition, dose, route of administration, patient population, and formulation that has been approved by the FDA in the United States.

**Ophthalmologist:** A doctor who specializes in diagnosing and treating eye and vision problems. Ophthalmologists can perform medical and surgical procedures relating to eye health and can manage various conditions that affect vision.

## P

**Patient acceptable symptom state (PASS):** The highest level of symptoms that a patient considers acceptable, as measured by a patient-reported outcome (PRO) scale. It’s a simple, two-part question that asks patients to assess their satisfaction with their current symptom state.

**Paramedic:** A specially trained medical technician licensed to provide a wide range of emergency services (such as defibrillation to restore a normal heartbeat or administering medications through an IV) before or during transportation to a hospital.

**Pathologist:** A doctor trained in identifying diseases by studying cells and tissues under a microscope.

**Pharmacologic remission:** A state in which a patient’s symptoms are significantly reduced or controlled through medication.

**Plasmapheresis (PLEX)/ Therapeutic plasma exchange (TPE):** A medical procedure that involves separating plasma (the liquid portion of the blood) from the rest of the bloodstream. The plasma is removed and replaced with purified fluid, such as healthy donor plasma, saline, or albumin. PLEX is used as treatment in some patients with MG because it helps to remove circulating antibodies that may contribute to the disease.

**Pediatric neurologist:** A doctor who specializes in diagnosing and treating neurological conditions in children.

**Peripheral fatigue:** Peripheral fatigue is weakness or tiredness in the muscles that results directly from muscle fatigue or problems at the neuromuscular junction.

**Pharmacist:** A licensed healthcare professional qualified to prepare and dispense medication. They play an important role in ensuring patients receive the right medications and understand how to use them safely.

**Physical therapist:** A healthcare professional trained to evaluate and treat people who have conditions or injuries that limit their ability to move or do physical activities.

**Post-Intervention Status (PIS):** The MGFA Post-Intervention Status is a tool used by clinicians to classify, or assess, how well a patient with MG responds to treatment. Status can be classified as either in remission (complete stable or pharmacological remission), minimal manifestations, or unchanged/worse. As a patient's clinical status and symptoms change over time, the Post-Intervention Status classification may change as well.

**Primary care physician (PCP):** A doctor in family medicine, general internal medicine, or general pediatrics who provides comprehensive care to the patients as their first point of contact and takes continuing responsibility for the patient's overall care.

**Ptosis:** Drooping eyelid(s).

**Pulmonologist:** A doctor who specializes in lung conditions, diagnosing and treating diseases of the respiratory system (includes the lungs, nose, throat, trachea, airways, muscles, and blood vessels involved in breathing). These doctors may be referred to as lung doctors, lung specialists, or chest doctors.

## Q

**Quantitative Myasthenia Gravis Score (QMG):** QMG is a test administered by a clinician to measure strength and fatigable weakness in patients with MG. The QMG score evaluates 13 areas, including eye muscles (ocular), swallowing muscles (bulbar), breathing muscles (respiratory), and muscles in the arms, legs, and trunk (limb/axial muscles). Special equipment is required to perform the QMG score. QMG scores range from 0 to 39, with lower scores indicating better muscle strength and function.

**Quantitative Myasthenia Gravis Revised (QMGr):** The QMGr is a newer version of the QMG score that also assesses muscle strength and fatigable weakness in the same areas. Like the QMG score, the QMGr is also administered by a clinician and requires special equipment. The QMGr has modified measurements for eyelid drooping and double vision to provide a more accurate assessment. QMGr scores range from 0 to 39, with lower scores indicating better muscle strength and function.

## R

**Receptor sites:** Structures located on the surface of cells that bind with specific proteins or chemicals. Receptor sites are often named after the signaling molecules they bind to, such as acetylcholine receptors. Acetylcholine receptors are found on muscle fibers and bind acetylcholine, which is released by nerves at the neuromuscular junction. When the acetylcholine attaches to its receptor, it causes a response in the cell. In this case, the binding of acetylcholine to its receptor causes a channel to open and sodium ions to flow into the muscle fiber, which is important for muscle contraction.

**Repetitive nerve stimulation (RNS):** A technique where a nerve is stimulated repeatedly and resulting muscle responses are recorded. RNS is used to evaluate how well signals are being transmitted by the neuromuscular junction and can help assess conditions like MG.

**Respiratory therapist:** A healthcare professional who is trained to assess and treat patients who have difficulty breathing.

**Risk Evaluation and Mitigation Strategy (REMS):** A drug safety program that the US Food and Drug Administration (FDA) can require for certain medications with serious safety concerns. REMS programs help to ensure that benefits of a medication outweigh its risks. REMS may include special requirements for how a drug is prescribed, dispensed, and monitored.

## S

**Serious adverse event (SAE):** Refers to any expected or unexpected undesirable event, related or unrelated to the therapy that is being studied, that results in any of the following outcomes: death, a life-threatening adverse event, inpatient hospitalization (not required as part of the treatment) or prolongation of existing hospitalization, a persistent or significant disability or incapacity, or cancer, or a congenital anomaly or birth defect.

**Seronegative myasthenia gravis:** Refers to individuals who exhibit myasthenia gravis symptoms but do not have detectable antibodies associated with MG (AChR, MuSK, or LRP4) in their blood. Researchers believe that this type of MG still derives from an autoimmune issue, but antibodies involved have simply not been identified yet. Approximately 5% of people with MG have seronegative MG.

**Side effect:** An undesired effect of a medication or medical treatment. These effects can vary in severity and might impact the patient's quality of life.

**Single-fiber electromyography (EMG):** A specialized test that allows a doctor to identify and record the electrical activity of individual muscle fibers. This technique is used to help diagnose disorders that involve neuromuscular transmission.

**siRNA (small interfering RNA):** A type of molecule that can turn off, or "silence," certain genes in the body. siRNA therapies are being studied in myasthenia gravis to block the production of specific proteins that play a role in the disease.

**Skeletal muscle:** Muscles that are connected to bones and allow a person to perform many movements and functions of the body. These are voluntary muscles controlled at will, and are the muscles affected by myasthenia gravis.

**Specialty pharmacy:** A type of pharmacy that dispenses specialty medications, including some that are used for treating myasthenia gravis. These pharmacies may also offer support services for patients.

**Speech language pathologist:** A healthcare professional who evaluates and treats people with communication and swallowing disorders. They can help improve speech, language, and swallowing abilities.

**Social worker:** Professionals who assist people in coping with challenges in their everyday lives, managing relationships, and solving personal and family problems. Social workers may specialize in serving a particular population or working in a specific setting, providing support and resources to improve overall well-being.

**Subcutaneous Immunoglobulin (SCIg):** A treatment that uses purified immunoglobulin from healthy donors. This treatment is given under the skin, instead of in the vein (IVIg).

**Symptom relapse:** The return or worsening of symptoms after a period of improvement or stability.

## T

**Thoracic surgeon:** A surgeon who specializes in the treatment of diseases and disorders of the chest, heart, and lungs. In the case of myasthenia gravis, these surgeons often perform a surgery called thymectomy.

**Thymectomy:** A surgery to remove the thymus gland.

**Thymic hyperplasia:** An increase in the number of cells in the thymus gland. Thymic hyperplasia is not considered a tumor or cancerous.

**Thymoma:** A tumor of the thymus gland. These tumors are most often benign. About 10% of patients with myasthenia gravis have thymomas, and conversely, about 40% of thymomas are associated with myasthenia gravis.

**Thymus gland:** A small gland located in the upper chest, just behind the breastbone (also known as the sternum). The main function of the thymus is to develop the immune system, including the ability of the immune system to recognize normal healthy cells in the body ("self") versus invading organisms like viruses and bacteria, or even transplanted organs ("non-self"). The thymus is especially important during childhood development, and its role gradually decreases after adolescence.

**Thymomatous myasthenia gravis:** A form of myasthenia gravis that is associated with the presence of thymoma. Thymomas are a rare form of cancer of the thymus gland. Thymomas are present in 10-15% of MG patients.

**T cells:** A type of white blood cell that plays many roles in the immune system. T cells work to destroy invading viruses and bacteria and regulate the immune system. Like B cells, they are produced in bone marrow and mature in organs such as the thymus. Synonyms: T lymphocyte, thymocyte.

**Tolerance:** The ability of the immune system to recognize the body's own cells as safe, and to not attack or destroy cells that are normal and healthy.

**Transient neonatal myasthenia gravis (TNMG):** TNMG is a type of MG that occurs in 10-15% of children born to mothers with MG. This happens when the mother's AChR antibodies cross the placenta to the baby. When born, infants with TNMG may have weakness, difficulty feeding, and difficulty breathing.

## V

**Voluntary muscles:** Muscles that are under voluntary control, meaning a person can choose to move or relax the muscle at will. Examples of voluntary muscles include the eye (extraocular) muscles as well as muscles in the face, neck, arms, and legs. In contrast, the heart is an example of an involuntary muscle because it works automatically and cannot be controlled by choice.