

Myasthenia Gravis and Immune Globulin Therapy

WHAT YOU NEED TO KNOW

Our goal is to make a positive difference in the lives of others! The patients and caregivers who give us the honor to improve their lives are members of the CSI family. We are passionate in our belief that all therapies should be individualized to meet the patient's specific needs. We want you to experience the difference and join our family today!

Contact us at 1-833-569-1005.





What Is Myasthenia Gravis (MG)

MG is a chronic autoimmune neuromuscular disease characterized by weakness of the skeletal muscles. Common symptoms include weakness of the muscles that control the eye and eyelid, facial expressions, chewing, talking, and swallowing. Weakness tends to increase during periods of activity and improve after periods of rest. The condition results from a defect in the transmission of nerve impulses to muscles, which is due to the presence of antibodies against acetylcholine. The exact reason this occurs is not known. Some cases have been linked to tumors in the thymus gland. Researchers believe that variations in certain genes may increase a person's risk to develop MG, but other factors likely also play a role. There is no cure for MG at this time, but treatment can significantly improve muscle weakness. Some cases may go into remission temporarily, and muscle weakness may disappear so that medications can be discontinued.

What are the symptoms?

MG Symptoms

MG causes muscle weakness and can affect any voluntary muscle. Voluntary muscles are the muscles that we directly control to make our body do things like walk, run, write, throw, lift, smile, and chew. In MG, the most commonly affected muscles are the muscles that control eye and eyelid movement, facial expressions, and swallowing. The degree of muscle weakness varies from person to person. People with more severe disease may have many muscles affected, including the muscles that control breathing. People with mild disease may have only one muscle group involved, such as the eye muscles.

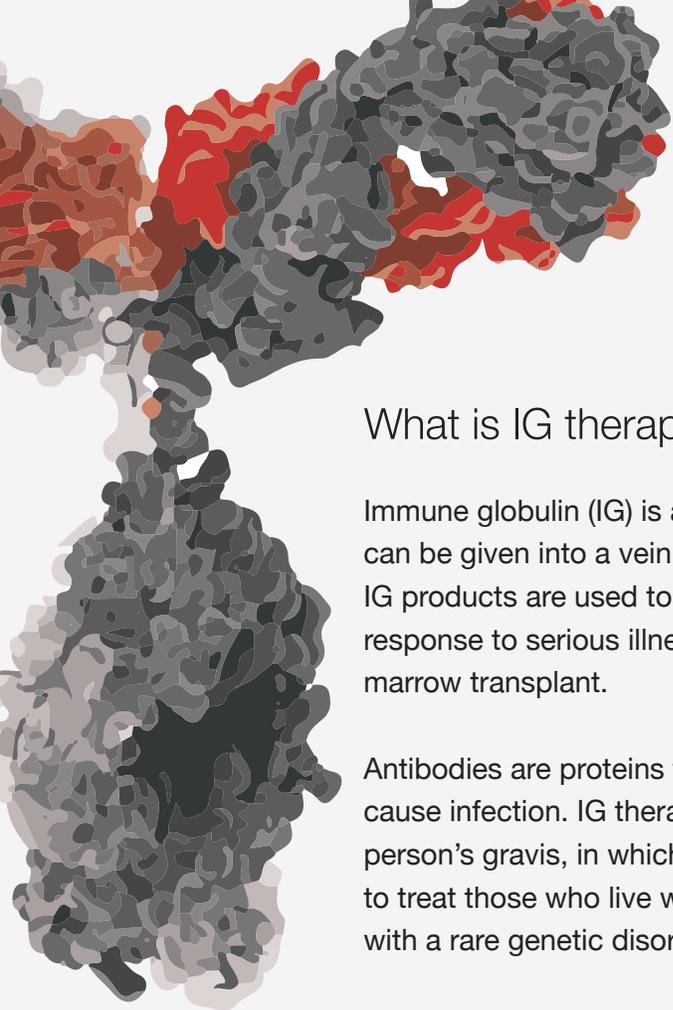
In general, signs and symptoms of MG may include:

- Drooping of one or both eyelids (ptosis)
- Blurred or double vision (diplopia)
- Unstable or waddling gait (manner of walking)
- Weakness in the arms, hands, fingers, legs, and neck
- Change in facial expression
- Difficulty swallowing and shortness of breath
- Impaired speech (dysarthria)

People can develop MG at any age. For unknown reasons, it is most commonly diagnosed in women younger than age 40 and men older than age 60. It is uncommon in children, but some infants born to women with MG have signs and symptoms of the condition for the first few days or weeks of life. This temporary occurrence of symptoms is called transient neonatal myasthenia gravis.

Questions & Answers

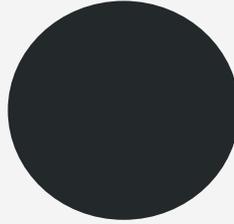
Immune Globulin (IG) Therapy



What is IG therapy?

Immune globulin (IG) is a sterile solution of concentrated human antibodies that can be given into a vein (intravenously or IV) or under the skin (subcutaneously or SC). IG products are used to treat disorders of the immune system, boost the immune response to serious illness, or treat immune suppression after someone has had a bone marrow transplant.

Antibodies are proteins that the body makes to help fight off bacteria and viruses that cause infection. IG therapy is used to treat autoimmune diseases, such as myasthenia person's gravis, in which the immune system is not functioning properly. It is also used to treat those who live with a dysfunctional immune system because they were born with a rare genetic disorder that affects the immune system.



How is IG made?

There is a highly specialized and lengthy process used to manufacture IG.

It begins in blood centers across the country where tens of thousands of healthy individuals donate their plasma (the portion of the blood where the immune globulins and other blood proteins are contained).

The plasma from all individuals is pooled together, then the immune globulins are separated and other blood proteins and harmful substances are removed. The result is a highly purified immune globulin preparation that is packaged and ready to be infused.



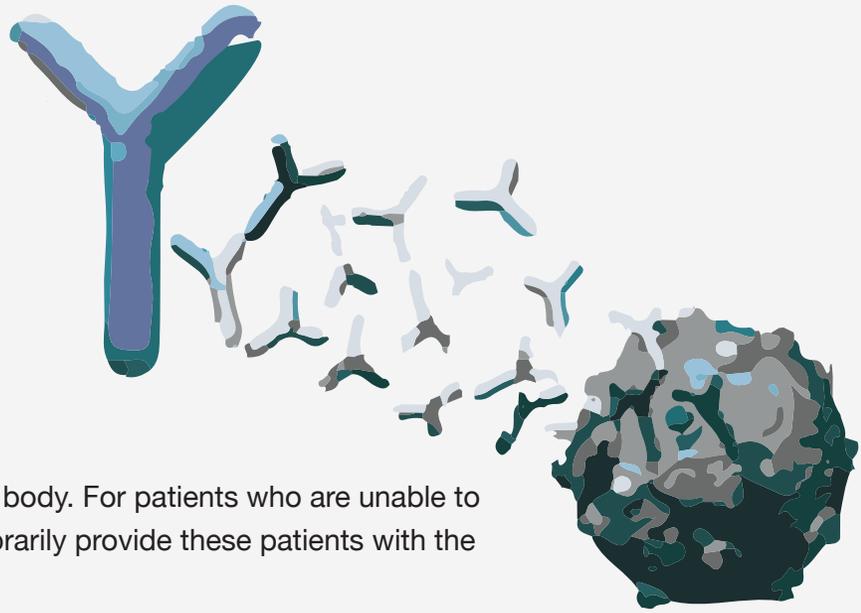
Is IG safe?

yes!

Since IG is derived from human plasma, theoretically there is a risk that viruses may be passed on to the recipient. However, every possible precaution and step is taken to ensure patient safety.

Blood donation centers are strictly monitored and regulated by the FDA, and IG manufacturers, as well as the individual donors, undergo an in-depth screening process to determine that they have not been exposed to certain pathogens, such as the HIV or hepatitis virus.

Furthermore, during the manufacturing process, steps are taken to inactivate and remove viruses as an extra precaution.

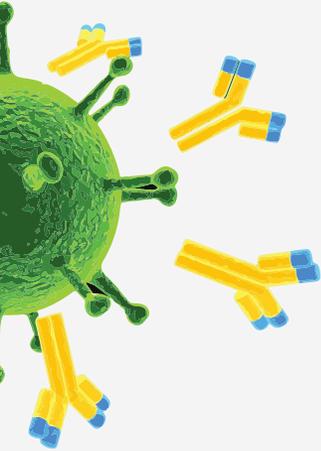


How does IG work?

It is unclear as to exactly how IG works within the body. For patients who are unable to produce their own antibodies, IG is used to temporarily provide these patients with the antibodies they need to ward off infection.

In patients with autoimmune diseases, or other conditions where the body's immune system is not functioning as it should, IG can help regulate an overactive immune system by signaling it to slow down or stop inflammatory processes.

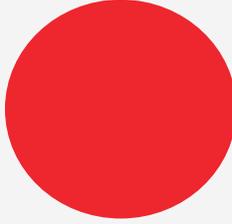
It is also possible that IG might redirect the out-of-control immune system from the body's tissues by serving as a target for the autoantibodies. While it is possible to debate the mechanisms by which IG successfully treats diseases, there is no denying its efficacy. IG has significantly improved the quality of life for many individuals.



Why do I have to have regular infusions?

IG does not affect your immune system's ability to produce immune globulins. It simply increases the antibody level within your body.

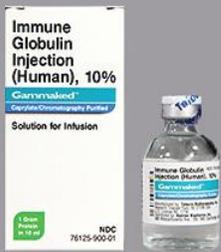
Antibodies, whether those produced naturally by your body or those infused in the form of IVIG or SCIG, are eventually metabolized and eliminated by your body, usually in about 3-4 weeks. Regular infusions are necessary to maintain immune globulin levels within the desired range .



What kinds of side effects can I expect?

The most common side effects include headache, nausea, low-grade fever, chills, rash, neck/back stiffness, and fatigue. Generally, these side effects are mild and tolerable and most often, they can be relieved by decreasing the rate of infusion. More serious side effects, such as allergic reactions, are rare, but have been reported. Should you develop an allergic reaction, your health care providers are sufficiently trained to handle this. Do not hesitate to contact your physician if your side effects are severe or persistent.

It is possible to reduce the severity of the side effects associated with IG infusions. Your physician, pharmacist, and nurse may suggest premedication with acetaminophen or antihistamines. Corticosteroids are also an option your doctor might consider. It is also important to make sure that you are sufficiently hydrated before your infusions, so be sure to drink plenty of water for several days before your infusion.



Are there different brands of IG? How are they different?

Currently there are eight brands of IVIG and six brands of SCIG available in the United States. Although each brand of IG is prepared through a unique process, they are all considered equal in terms of clinical effectiveness.

Different brands vary in terms of formulation, and have different side effect profiles that can also be patient-specific. Product characteristics that can affect a patient's ability to tolerate infusions include, how concentrated the solution is, how much sugar or salt it contains, how much of a particular immune globulin (IgA) it contains, and how acidic the solution is.

Your physician and pharmacist will examine these factors and identify the best brand for you.



Is immune globulin therapy covered by insurance for myasthenia gravis?

Most health insurance companies cover IG therapy for MG under certain conditions. These criteria vary according to each plan, and all require documentation to support the medical need. In general, immune globulin is considered medically necessary for the treatment of myasthenic exacerbation when all of the following criteria are met:

- a. Diagnosis of generalized myasthenia gravis **AND**
- b. Evidence of myasthenic exacerbation, defined by at least one of the following symptoms in the last month:
 - (1) Difficulty swallowing
 - (2) Acute respiratory failure
 - (3) Major functional disability responsible for the discontinuation of physical activity
 - (4) Recent immunotherapy treatment with a checkpoint inhibitor [e.g., Keytruda (pembrolizumab), Opdivo (nivolumab), Tecentriq (atezolizumab)] **AND**
- c. One of the following:
 - (1) History of failure, contraindication, or intolerance to immunomodulator therapy (e.g., azathioprine, mycophenolate mofetil, cyclosporine, methotrexate, tacrolimus) for long-term management of myasthenia gravis.
 - (2) Currently receiving immunomodulator therapy (e.g., azathioprine, mycophenolate mofetil, cyclosporine, methotrexate, tacrolimus) for long-term management of myasthenia gravis **AND**
- d. IVIG dose does not exceed 2,000 mg/kg per month given over 2 to 5 days administered in up to three monthly infusions. Dosing interval may need to be adjusted in patients with severe comorbidities.

How can CSI Pharmacy help you?

Clinical Specialty Infusions (CSI) Pharmacy is a nationwide specialty pharmacy dedicated to servicing patients with chronic and rare illnesses requiring complex care. We are invested in improving the health-and the lives-of the people we serve by offering expert clinicians, passionate advocates, committed healthcare partners, and strategic reimbursement experts. Our goal is to optimize patient outcomes, while treating patients as if they were members of our family.

CSI Pharmacy's commitment to the MG community

We feel honored and privileged to be able to play an important role in helping patients and their families navigate their healthcare needs. Our mission is to improve quality of life while providing accurate education about the disease and treatments available. Our team, which includes patient advocates, nurses, and pharmacists, are readily available to the patient and their caregivers. As a commitment to our patients, CSI Pharmacy plays an active role in rare disease patient organizations, including national, state, and local communities and support groups. We enjoy sponsoring, endorsing, and participating in the programs of the Myasthenia Gravis Foundation of America (MGFA) and the Myasthenia Gravis Hope Foundation.

CSI Pharmacy is committed to helping those in need, *whether or not you are our patient*. Please let us know if we can be of help to you.

CSI Ig **Promise** Program:

With CSI Pharmacy, it is our team members that make the difference! Our team created a program tailored to the specific needs of a patient undergoing Immune Globulin Therapy. We call this program our **CSI Ig PROMISE!**

We **Promise** to provide the following services:

- A dedicated **Intake Specialist** who is thoroughly trained in understanding and accessing health insurance benefits. Every insurance company has its own coverage policy regarding IVIG/SCIG for its beneficiaries. Our specialists will work with you and your insurance company to be sure you receive the appropriate coverage for your specific needs.
- Every patient will be assigned a dedicated **Patient Advocate** who will:
 - Help you navigate and coordinate your course of treatment and assist in making sure your healthcare needs are met.
 - Assist with navigating the complexities of health insurance and ensure that your plan is affordable and appropriate for your health care needs.
 - If necessary your advocate will provide information on Health Insurance Co-Pay assistance and other programs available for your specific needs.
- A dedicated **Clinical Pharmacist and Infusion Nurse**
 - The pharmacist and nurse are specialized, experienced, and certified in immune globulin therapy.
 - They collaborate in managing your infusion and customize it to your unique needs (product selection, labs, infusion rate, adverse events, clinical response).
 - Your Infusion Nurse is with you from the beginning to the end of your infusion.

Your Infusion Pharmacist, Infusion Nurse, and Patient Advocate are on call 24/7/365, so they are always available in the event you need them or have questions.

CSI Pharmacy ensures that your therapies are accessible and affordable

CSI Pharmacy has trained individuals who perform a complete benefit investigation to find out about medication coverage through your insurance plan and any copay or out-of-pocket costs that may be associated with your coverage.

- 1 . We will provide you with access to manufacturer copay programs, where eligible.
2. If you are uninsured or underinsured, we will assist you in navigating through our complicated healthcare system to determine all options to purchase affordable health insurance. If that is not possible, we will share with you ALL resources available through foundations, other assistance programs, and possibly Social Security Disability, where eligible.

Let CSI Pharmacy help you concentrate on the things that matter: getting better. We will help you understand your medication through educational and clinical programs and provide resources from caring professionals, who take pride in assisting you and your loved ones.



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CSI Pharmacy is a nationwide pharmacy serving patients in all regions of the United States. Please visit our website to see a list of the states in which we are currently located, as well as those in which we will be licensed soon.