



# Your Questions Answered

## MYASTHENIA GRAVIS

**Q** *Is plasmapheresis an effective treatment for myasthenia gravis?*



**DR. JULIE ROWIN RESPONDS:**

**A** Several treatment options are available for people with myasthenia gravis (MG). In my experience, plasmapheresis (plasma exchange) has been the most effective in treating the severe symptoms of an MG crisis, such as difficulty breathing or swallowing. But while plasmapheresis reverses severe symptoms of MG in an exacerbation quickly, the effect can be short-lived.

A rare autoimmune disease, MG causes weakness in the voluntary muscles. In people with MG, these muscles rapidly grow weaker with repeated use (and then recover after a period of rest). Myasthenia gravis is caused by an abnormal antibody in the blood that prevents a substance in nerve cells, called acetylcholine, from doing its job. This neurotransmitter communicates with muscles, telling them to contract. The muscles affected by MG often involve those that control eye and eyelid movement, vision, swallowing, chewing, and breathing. With treatment, many people with MG improve significantly.

During plasmapheresis, the fluid part of the blood (plasma) containing the abnormal antibodies is removed and replaced with other fluids before being returned to the patient. Since the effects are often short-term (generally two weeks), a series of treatments over a period of weeks is usually necessary, typically on an outpatient basis. The process can be costly, and potential risks exist, such as a dangerous drop in blood pressure, bleeding as a result of an-

ticoagulants used with the procedure, and possible infection in cases where a peripheral vein (a vein not in the chest or abdomen) cannot be used and a catheter must be inserted into the chest.

Intravenous immunoglobulin (IVIG) is another treatment used to treat serious or worsening cases of MG. Rather than removing abnormal antibodies, IVIG involves the injection of serum containing antibodies in order to affect the immune system's own antibody production. Treatments last three to four weeks and are done by IV through a peripheral vein. For the most part, side effects are mild and may include chills and headache during and after infusion, although some patients may be at risk of serious adverse events such as anaphylactic shock or kidney failure.

Plasmapheresis and IVIG have been shown to be comparable in clinical trials. Whereas plasmapheresis has yet to be compared to placebo in clinical trials, IVIG has been compared to placebo and been shown to be effective in MG, particularly in moderate disease. Most experts believe that plasmapheresis is more potent and reliable in treating patients who are in crisis. When choosing between the two treatments, several factors are considered, including severity of the symptoms and potential side effects for the individual. Whichever treatment you choose, the decision should be made in the context of a long-term strategy to keep the MG under control.



**Ptosis (drooping of the eyelid) is one of the common symptoms of myasthenia gravis.**

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**MOYAMOYA**

**Q** What treatments are available for Moyamoya disease?



**DR. MARIO  
ZUCCARELLO  
RESPONDS:**

**A** A number of treatment options are available for people with Moyamoya disease (MMD). The best course of treatment will be determined by the patient's age, his or her symptoms, and the size of the stroke caused by MMD.

The disease is caused by blocked arteries at the base of the brain. To compensate for the narrowing arteries and facilitate the flow of oxygen-rich blood, the brain creates collateral blood vessels. When seen on a brain scan, these tiny blood vessels have a hazy, filmy appearance—the “puff of smoke” that gives the disorder its Japanese name. The narrowing of these collateral blood vessels can lead to stroke.

The chance of being diagnosed with MMD in the United States is about one in a hundred thousand. While the cause of MMD is not completely known, a genetic component is suspected of playing a role, particularly in Asian populations. Also, several diseases—including sickle cell anemia, Down syndrome, and neurofibromatosis—have been associated with MMD and may be potential causes.

Once a diagnosis of MMD is confirmed, several treatments may be considered. If few symptoms have appeared, then an anti-platelet drug such as aspirin or clopidogrel (Plavix) might be prescribed to inhibit clot formation.

If there has been a transient ischemic attack (TIA), which is when blood flow to a part of the brain stops for a brief period of time, then revascularization is recommended to prevent future stroke. In direct revascularization, blood vessels outside the brain are connected to blood vessels inside the brain to create immediate blood flow, which increases the amount of oxygen and other nutrients delivered to the brain. During indirect revascularization, an artery or muscle from the forehead is placed on the brain's surface with the hope that, over time, new blood vessels will develop and increase circulation.

While the outlook for someone with MMD depends upon the how rapidly the blockage occurs and by the overall neurologic condition of the patient, revascularization surgery is quite effective in decreasing the progression of strokes. Left untreated, approximately half of MMD patients will continue to experience strokes, seizures, headaches, TIAs, and impaired day-to-day function such as compromised speech or motor function.

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**RADIATION**

**Q** What are the effects of radiation therapy on the brain and spine?



**DRS. ORLY AVITZUR  
AND LYNNE TAYLOR  
RESPOND:**

**A** Often used in the treatment of tumors, radiation therapy (RT) can injure the healthy brain and spinal cord tissue nearby. It's believed that such injury is caused by damage to the blood vessels that supply these areas of the body and damage to the glia, which are the supportive cells of the central nervous system.

The neurologic complications caused by RT may occur immediately or weeks, months, or years after treatment. Five percent of patients experience early or acute injury, which occurs during the first two weeks after treatment. Symptoms include headache, nausea and vomiting, sleepiness, and fever.

Fifty percent of patients, especially children, experience early-delayed injury, which occurs from weeks to several months following RT. Symptoms include drowsiness, fatigue, and cognitive problems such as difficulty with thinking and memory. These changes are reversible and typically respond to treatment with steroids.

Late-delayed injury can follow many months or longer after treatment and is associated with permanent white matter changes in the brain, loss of brain tissue, and—if severe—dementia and death. These effects will vary based on other treatments or underlying diagnoses but can be present in up to 50 percent of people treated with whole brain radiotherapy and chemotherapy for central nervous system lymphoma.

When RT-induced injury occurs in the spinal cord, patients may feel a loss of sensation and weakness in the legs and experience bowel and bladder difficulties. Early-delayed damage to the spinal cord is often reversible, whereas late-delayed is often permanent. The risk of developing late-delayed injury increases with radiation dose, age, conditions such as diabetes and high blood pressure, and when RT is combined with chemotherapy.

To avoid long-term consequences of RT, patients should see a neuro-oncologist or a radiation therapist with expertise in neuro-oncology.

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