

What is ocular myasthenia gravis?

Ocular myasthenia gravis is a form of myasthenia gravis (MG) in which the muscles that move the eyes and control the eyelids are easily fatigued and weakened.

What are the common symptoms of ocular myasthenia gravis?

People with ocular MG have trouble with sight due to double vision and/or drooping eyelids. Their eyes do not move together in balanced alignment, causing them to see “double” images. One or both eyelids may droop to cover all or part of the pupil of the eye, thus obstructing vision.

These symptoms may be mild to severe. Eye weakness often changes from day to day and over the course of a day. Problems with the eyes are often worse at the end of the day or after the eyes have been used for a prolonged period of time. Many people with ocular MG find that their eye problems are temporarily improved if the eyes are rested by closing them for several minutes when symptoms are troubling.

People with ocular MG do not have difficulty swallowing, speaking or breathing, nor do they have weakness of the arms or legs. Descriptions of the symptoms that people with ocular MG can have include:

The MGFA mission is to facilitate the timely diagnosis and optimal care of individuals affected by myasthenia gravis and closely related disorders and to improve their lives through programs of patient services, public information, medical research, professional education, advocacy and patient care.

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355 Lexington Avenue, 15th Floor
New York, NY 10017-6603

(800) 541-5454

(212) 297-2156 • (212) 370-9047 fax

mgfa@myasthenia.org • www.myasthenia.org

www.myasthenia.org
800.541.5454

Ocular Myasthenia Gravis

M G

**Common questions about
Ocular Myasthenia Gravis.**

F A

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• **Double vision** –

Seeing two images rather than one. This results from weakness of the muscles that move the eyes together in alignment. The medical term for double vision is diplopia. Some people experience blurred vision rather than double vision when the eyes are not properly aligned.

• **Drooping eyelids** – The eyes do not appear to be opened fully. If the eyelid covers the pupil of the eye, then the vision of that eye will be obstructed. The medical term for drooping eyelids is ptosis (pronounced “toe-sis”).

Who gets ocular myasthenia gravis?

Problems with double vision and drooping eyelids are often the first symptoms of MG. Although most people with MG have eye problems at disease onset, most will go on to develop weakness in other muscles—a condition referred to as generalized myasthenia gravis. This typically occurs within the first two years after the initial diagnosis. About one in six or seven people with MG (15%) will have only ocular problems, i.e. ocular MG.

Among the people with MG who have only ocular problems during their first year of illness, about half will go on to develop more generalized muscle weakness. People who have had only ocular MG for five years will probably not develop additional MG weakness.

It is not possible to predict which patients will have ocular MG and which will have more generalized MG symptoms. Most people with congenital

(non-autoimmune) MG do not have ocular MG. People with ocular MG are slightly more likely to have seronegative MG (no measurable acetylcholine receptor antibodies) compared with people with generalized MG.

Why are the eye muscles frequently involved in myasthenia gravis?

There may be several reasons why eye muscles are more frequently involved. However, this is not completely understood.

One hypothesis is that patients may simply notice eye weakness more often than mild weakness in other muscle groups in the body. Another hypothesis is that the eye and eyelid muscles are structurally different from muscles in the trunk and limbs. For example, they have fewer acetylcholine receptors, which is where the defect occurs in autoimmune MG. Eye muscles contract much more rapidly than other muscles and may be more likely to fatigue.

Perhaps the most important difference between eye and eyelid muscles compared with other muscles of the body is that eye muscles respond differently to immune attack. The differences in the response of eye muscles to immune attack may explain why eye muscles are also targeted in other autoimmune conditions, such as autoimmune thyroid disease.

How is ocular myasthenia gravis treated?

People with ocular MG and their caregivers should balance the severity of the symptoms with the risks and benefits of treatment. People who have primarily cosmetic problems due to ptosis or diplopia may consider nonpharmacological treatment, such as:

• Wearing dark glasses in bright light, which some patients find helpful.

- Using eyelid tape (a special type of tape used to hold the eyelids open without injuring the eyelids). This can be used for ptosis and may be preferable to drug therapy that alters the immune system using agents such as glucocorticoids (prednisone or similar agents), azathioprine (Imuran[®]), cyclosporine or mycophenolate mofetil (CellCept[®]).
- Applying a patch to one eye. This permits patients with double vision to see one image. If the same eye is consistently patched, vision in that eye will decrease. Therefore, it is important to alternate the patch from one eye to the other to avoid permanent vision loss.
- Using eyelid crutches (clever devices attached to glasses to hold the eyelids open) for ptosis and eyeglass prisms for diplopia. These are rarely-used, older methods of treatment for ocular MG.

When ocular symptoms are severe or disabling, treatment with immune system modulating therapy may be considered.

Agents that improve neuromuscular transmission, such as Mestinon[®], may be helpful for ptosis, but are generally not very useful for diplopia.

Thymectomy is usually not considered for people with ocular MG unless the manifestations are severe or disabling.

Eyelid or eye muscle surgery is generally contraindicated in MG patients.

Medications such as prednisone or steroid-sparing agents such as azathioprine or mycophenolate mofetil may be prescribed, and is usually done on a case-by-case basis taking both potential risks and benefits into account.