

Myasthenia Gravis

What is myasthenia gravis?

Myasthenia gravis is an autoimmune disease in which antibodies attack and weaken normal skeletal muscle tissue. It occurs in both children and adults and can affect different muscle groups in the body. Symptoms include double vision, droopy eyelids, easy muscular fatigue and breathing/swallowing difficulty. The symptoms tend to worsen with increased activity and improve with rest. The eyelid and extra-ocular muscles are commonly involved in myasthenia, either in isolation (ocular myasthenia gravis) or in conjunction with other skeletal muscles (systemic myasthenia gravis)[See figure 1].



Fig. 1: Droopy left upper eyelid.

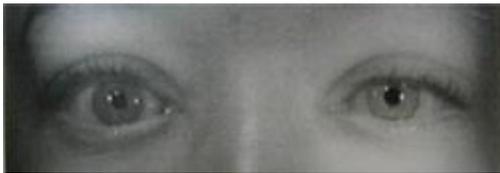


Fig. 2: Left upper eyelid elevated during Tensilon test.

What causes myasthenia gravis?

The condition develops when auto-antibodies attack normal skeletal muscle tissue. Nerve endings that innervate skeletal muscle typically release a chemical, acetylcholine, to induce the muscles to contract. In myasthenia, antibodies attach themselves to the receptors for the acetylcholine on the skeletal muscle. Thus, when acetylcholine is released from a nerve and tries to induce contraction in a muscle, the receptors cannot attach to the acetylcholine resulting in muscle weakness and fatigue.

How common is myasthenia gravis?



Myasthenia is an uncommon disease. The incidence is estimated at 20/100,000. There appears to be no racial or sex predilection in myasthenia. It is not a directly hereditary condition.

What are the characteristic symptoms in myasthenia gravis?

Approximately 90% of people with myasthenia gravis have ocular involvement and 50% present with ocular symptoms. Double vision from eye misalignment ([strabismus](#)) and drooping of the eyelids ([ptosis](#)) are the most common symptoms. These symptoms classically vary and worsen throughout the day. The double vision/ptosis may be present in the evening while not present in the morning. Less common symptoms are facial weakness, chewing and swallowing difficulty, respiratory weakness, and arm and leg muscle fatigue.

How is myasthenia gravis diagnosed?

The diagnosis of myasthenia can be challenging. No one test is positive 100% of the time.

Examination findings are an important tool to make the diagnosis. Ice testing (placing ice on a closed ptotic, or droopy, eyelid) is another way to diagnose myasthenia and can sometimes be done in your ophthalmologist's office. Most other testing such as administration of medications like Tensilon or Prostigmin (see figure 2), or electromyography (EMG) are most often performed by a neurologist. The ice test, Tensilon test, and EMG may be difficult to perform in young children due to cooperativity. Laboratory testing such as acetylcholine receptor antibody panels can be done but are unfortunately positive in only about 50% of patients with ocular myasthenia.

What other conditions can occur in association with myasthenia gravis?

Myasthenia can occur in association with other autoimmune diseases, most commonly thyroid disease, rheumatoid arthritis, and diabetes. A thymoma (benign tumor of the thymus) in the chest cavity is another association, although very rare in children. Chest imaging such as an X-ray or CT scan may be ordered to assess the thymus in patients with myasthenia gravis.

How is myasthenia gravis treated?

Myasthenia is rarely cured, but the symptoms can be improved with a number of different medical or surgical interventions. The main goal of treatment from a biochemical perspective is to increase the amount of acetylcholine available to the receptors on the nerve endings in the muscle. This can be done either by increasing the amount of the acetylcholine itself by minimizing its breakdown, or by destroying the antibodies that are attacking the acetylcholine receptors. Mestinon, or pyridostigmine, is a cholinesterase inhibitor which blocks the breakdown of acetylcholine. This is a commonly used medication to treat the symptoms of myasthenia, but it has been demonstrated that this medication is more effective at alleviating the non-ocular symptoms than the ocular ones. Immunosuppressive agents such as prednisone can be effective at blocking the immune response involved in myasthenia. Plasmapheresis is a procedure whereby the offending antibodies are removed directly from the bloodstream. This treatment



frequently needs to be repeated as symptoms can return after time. IVIG is an intravenous treatment that affects the production and/or function of the abnormal antibodies. In some cases surgical removal of a thymoma (or normal thymus tissue in children) can improve symptoms. Severe cases of myasthenia that do not respond to the aforementioned treatments may require targeted immune-based therapy to suppress the immune system, including medications such as rituximab.

When should eye surgery be considered?

The ptosis and strabismus related to myasthenia can at times be helped with surgery. Surgical intervention is sometimes advised if the ptosis and/or strabismus deviation has been stable for at least one to two years. Surgeons are typically conservative when planning to operate on patients with myasthenia due to possible complications of surgery such as severe eye dryness and worsening eye misalignment or double vision

How can I find out more information about myasthenia gravis?

- Contact the [Myasthenia Gravis Foundation of America, Inc.](#)

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