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## Myasthenia gravis

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### Summary

- Myasthenia gravis is an autoimmune disease that causes muscle weakness.
  - The symptoms are caused by the immune system interfering with the transmission of messages from the nervous system to the muscles.
  - There is no cure, but the symptoms can be managed.
  - Treatment options include drugs to suppress the activity of the immune system, plasmapheresis to clear the antibodies from the blood and surgical removal of the thymus gland.
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The term 'myasthenia gravis' (MG) comes from the Greek word 'myasthenia' meaning muscle weakness and the Latin word 'gravis' meaning severe. It is an autoimmune condition that causes problems with the transmission of signals from the nerves to the muscles. This results in weak muscles that get tired quickly and which improve after rest.

In the early stages, myasthenia gravis mostly affects the muscles that control eye movement, facial expression, chewing and swallowing. As the condition progresses, neck and limb muscles may also be affected, causing difficulty with holding the head up, walking upstairs and raising the arms. If untreated, breathing may be affected. Fortunately, treatment – which may include medication or surgery – is usually successful in managing the symptoms of the condition.

Myasthenia gravis affects all races and can develop at any age from childhood to old age. Women are affected nearly three times more often than men during early adulthood (under 40 years of age). After 50 years of age, more men are affected than women. It is fairly unusual for children under the age of 15 to have myasthenia gravis, except in some Asian countries where up to half of people with myasthenia gravis have symptoms beginning in childhood.

Although the condition doesn't generally run in families, people who inherit a tendency to develop autoimmune conditions are at increased risk of developing myasthenia gravis, so a person with myasthenia gravis may have another autoimmune disease, such as diabetes, or have a relative with an autoimmune disease.

A 2009 survey found that 2,574 people in Australia were currently being treated for myasthenia gravis. This equals approximately 1.2 out of every 10,000 people.

### Symptoms of myasthenia gravis

Some of the symptoms of myasthenia gravis include:

- weak muscles
- weakness that improves after resting and gets worse after physical activity
- visual disturbances such as double vision, inability to hold a steady gaze and droopy eyelids
- fatigue
- swallowing difficulties
- breathing difficulties
- shortness of breath.

### Cause of myasthenia gravis

Myasthenia gravis is just one of many autoimmune diseases, which include arthritis and type 1 diabetes. Normally, the immune system produces antibodies that recognise foreign things that enter the body, such as bacteria and viruses. This leads to them being destroyed and cleared from the body. In the case of an autoimmune condition,

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the body's immune system produces antibodies against things in the body that aren't foreign. In myasthenia gravis it is the structure at the junction of the nerves and the muscles (the neuromuscular junction) that is attacked.

About 85 percent of patients with myasthenia gravis produce antibodies against a protein called the 'acetylcholine receptor' (AChR). This is found at the neuromuscular junction and acts as a receiver for the chemical signal 'acetylcholine' (ACh) that is released from the nerve to tell a muscle to contract.

The antibodies bind to the acetylcholine receptors on the surface of the muscle and greatly reduce their ability to receive the chemical signal. As a result, the person experiences muscle weakness, which becomes worse as they repeatedly try to use the same muscle.

Many people with myasthenia gravis who don't have antibodies to the AChR, have antibodies to a protein called 'muscle-specific kinase' (MuSK). This protein helps organise ACh receptors on the muscle cell surface. Research is ongoing to find out what antibody is responsible in the approximately 10 percent of people who don't have antibodies to AChR or MuSK. Recently, antibodies to a protein called 'LRP4' were found to be the cause for some of these people.

Scientists don't know what triggers most autoimmune conditions, but they have a few theories. One possibility is that certain viral or bacterial proteins mimic 'self-proteins' in the body (such as AChR), stimulating the immune system to accidentally attack it.

There is also evidence that the thymus gland plays a role in myasthenia gravis. About 15 percent of people with myasthenia gravis have a thymic tumour, called a thymoma, and another 65 percent have an overactive thymus, a condition called thymic hyperplasia. When the thymus doesn't work properly, the immune system might lose some of its ability to distinguish self from non-self, making it more likely to attack the body's own cells.

### **Myasthenia gravis crisis**

A crisis occurs when the muscles involved in respiration are affected. Symptoms include shortness of breath and breathing problems. This is a medical emergency that needs hospitalisation and prompt medical treatment, including the use of a ventilator to assist breathing.

Some of the triggers of myasthenic crisis include physical stress, pregnancy or infection. When myasthenia is properly treated, crisis is very rare. And when crisis does occur, it has a good rate of recovery, thanks to a wide range of treatments and the quality of respiratory care at most hospitals.

### **Diagnosis of myasthenia gravis**

If a person's physical examination and medical history reveals a pattern of weakness that suggests myasthenia gravis, further tests done to confirm the diagnosis include:

- A blood test can detect the presence of antibodies to the acetylcholine receptor (AChR) or MuSK. The majority of people with myasthenia gravis have antibodies to one of these proteins and this confirms the diagnosis of myasthenia gravis.
- Electromyography (EMG) uses electrodes to stimulate muscles and evaluate muscle function. Muscle contractions that become progressively weaker may indicate myasthenia gravis.
- The 'Tensilon test' is often used to diagnose myasthenia gravis. It involves an injection of a drug called Tensilon, which temporarily improves muscle strength in people with myasthenia gravis.
- Chest x-ray, CT scan or MRI may be performed to examine the thymus gland, because abnormalities of the thymus are often linked with myasthenia gravis.

### **Treatment for myasthenia gravis**

There is no cure for myasthenia gravis, but the symptoms can be managed. Some of the treatment options include:

- **changes to lifestyle** – such as getting adequate rest every day and minimising physical exertion
- **anti-acetylcholinesterase agents** – the most commonly prescribed anticholinesterase medication is pyridostigmine (Mestinon). These drugs prevent ACh destruction and increase the accumulation of ACh at neuromuscular junctions, improving the ability of the muscles to contract. The benefits of Pyridostigmine occur

within 30 to 60 minutes, but wear off in three to four hours, so tablets should be taken at regular intervals throughout the day. Side effects include excessive salivation, involuntary muscle twitching (fasciculation), abdominal pain, nausea, and diarrhoea. Other drugs may be used with anticholinesterase medications to reduce gastrointestinal side effects

- **corticosteroids** – (such as prednisone) suppress the antibodies that interfere with the function of the neuromuscular junction and may be used in conjunction with anticholinesterase medication. Corticosteroids improve symptoms within a few weeks and once improvement stabilises, the dose is slowly decreased. A low dosage may be used indefinitely to treat myasthenia gravis. However, side effects such as gastric ulcers, osteoporosis (bone thinning), weight gain, high blood sugar, and increased risk of infection may develop over the long term
- **other immunosuppressive drugs** – such as azathioprine and mycophenolate mofetil (CellCept) may be effective with fewer side effects than corticosteroids. Cyclophosphamide and cyclosporin are considered effective against myasthenia gravis, but because of their side effects they are only prescribed as a last resort when other medications are not effective
- **plasmapheresis** – blood is taken out and the acetylcholine receptor antibodies are removed. The ‘cleaned’ blood is then returned to the person’s body. The results only last a few weeks, as the immune system continues to create the antibodies. Plasmapheresis is often used to help resolve a myasthenic crisis. In severe cases, a person may need plasmapheresis as a long-term treatment
- **intravenous immunoglobulin** – is a short-term management strategy that involves an injection of nonspecific antibody (immunoglobulin), which temporarily stops the immune system’s production of its own antibodies
- **surgical thymectomy** – surgical removal of the thymus gland is recommended for some people. The benefits of thymectomy develop gradually and most improvement occurs years after the procedure is performed, but it is believed to be the only treatment capable of producing long-term, drug-free remission.

#### Where to get help

- Your **GP (doctor)**
- **Muscular Dystrophy Australia** Tel. **(03) 9320 9555**

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