

## Myasthenia gravis

Myasthenia gravis is an autoimmune disease characterised by muscular weakness. In order to move at will, voluntary muscles receive chemical messages (neurotransmitters) from nerves (motor neurones) at special structures called neuromuscular junctions, where the nerve joins the muscle fibre. The neurotransmitter passes to a receptor, which is activated and causes the muscle to contract.

In myasthenia gravis, the immune system stops the action of the neurotransmitter by blocking or destroying the receptor in the neuromuscular junction - this prevents the muscle from contracting. This blocking of the chemical messages causes the muscular weakness. Usually, a person affected by this disease has a worsening of symptoms after physical exertion and feels better after resting. The disease usually targets the eyes first. Sometimes, the disease progresses further and interferes with swallowing, chewing and talking. Myasthenia gravis is rarely fatal unless the muscles of respiration are impaired. Around three in every 10,000 people are thought to have the disease. Women are twice as likely as men to develop myasthenia gravis, although anyone of any age or either sex can have the disease. There is no cure, but treatment can ease the symptoms.

### Symptoms of myasthenia gravis

Some of the symptoms of myasthenia gravis include:

- Weak muscles
- The weakness 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.

### Communication between nerves and muscles

The nervous system relays messages to muscles via a special chemical. The nerve cells use the chemical acetylcholine to transmit messages to the receptor of the muscle cell, much like a key fitting into a lock. The received message is then acted upon - for example, the muscle cell contracts. In myasthenia gravis, the immune system creates antibodies that attack the receptors of the muscle cells so that the chemical 'key' can no longer fit. Vigorous muscular activity requires increased chemical messages. This helps to explain why people with myasthenia gravis have a worsening of symptoms after physical exertion and feel better after resting.

### Cause of myasthenia gravis

Researchers are confident that myasthenia gravis is an autoimmune disorder, but the cause of this abnormal immune response is not known. In some people with myasthenia gravis, the thymus gland seems to be involved. The thymus, located in the chest, is a structure of the immune system. A baby has a disproportionately large thymus compared to an adult, because the gland shrinks as a person matures. However, a person with myasthenia gravis typically has an enlarged thymus. Around 10 to 15 per cent also have abnormalities such as tumours of the thymus (thymomas), which may turn cancerous in some cases. The exact role of the thymus in myasthenia gravis isn't known, but this gland is clearly associated with the production of acetylcholine receptor antibodies.

### The 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.

## Diagnosis of myasthenia gravis

Myasthenia gravis is diagnosed using a number of tests including:

- **The Tensilon test** - a drug (edrophonium) given intravenously. If the person's muscle strength temporarily improves, this indicates myasthenia gravis.
- **Electromyography (EMG)** - a device that measures muscular response to nerve stimulation. A person with myasthenia gravis has an atypical response to the EMG test.
- **Blood test** - to check for the presence of acetylcholine receptor antibodies.

## 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** - various drugs can modify the presence or activity of acetylcholine. This helps to increase the number of messages between the nerve cells and muscle cells. Possible side effects of treatment include allergies, interaction with other drugs, and overdose symptoms such as diarrhoea, nausea and vomiting.
- **Corticosteroids** - these drugs work by suppressing the activity of the immune system and reducing the production of acetylcholine receptor antibodies. It is suspected that corticosteroids may also prompt the body to build more acetylcholine receptors in the muscle cells. Despite the good results, corticosteroids are more likely than any other type of therapy to cause unwanted side effects.
- **Other immunosuppressive drugs** - such as azathioprine and cyclosporin.
- **Plasmapheresis** - blood is taken out and the acetylcholine receptor antibodies 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** - a short term management strategy that involves the intravenous administration of immunoglobulin. However, this treatment is quite expensive.
- **Surgical thymectomy** - surgical removal of the thymus gland. It is unclear how surgical thymectomy leads to a remission of symptoms, but research shows that a person's level of acetylcholine receptor antibodies drops after surgery.

## Where to get help

- Your doctor
- Muscular Dystrophy Association Tel. (03) 9320 9555

## Things to remember

- Myasthenia gravis is an autoimmune disease characterised by muscular 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.

**This page has been produced in consultation with, and approved by:**

Muscular Dystrophy Association

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