NHS
Greater Glasgow and Clyde

Information about

Myasthenia Gravis

West of Scotland Myasthenia Service
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What is Myasthenia Gravis?

For our muscles to work, the brain sends an electrical message to the **nerve endings**. The nerve endings release a chemical called a **neurotransmitter (acetylcholine)** into the **neuromuscular** junction (this is a tiny gap between the nerve endings and the surface of the muscle fibres). The acetylcholine quickly attaches to the receptors on the muscle fibres. This, in turn, triggers the muscle fibres to contract (move).

People with Myasthenia Gravis have a fault in the way electrical messages travel from the nerves to the muscles. As a result of this defect, the muscles are not stimulated properly. So the muscles easily tire and become weak.

Myasthenia Gravis affects the skeletal muscles in the body. Skeletal muscles are the ones attached to bones. Muscles that can be affected in Myasthenia Gravis include:

- Those which move the arms and legs
- The face and eye muscles
- Muscles involved with swallowing and breathing.

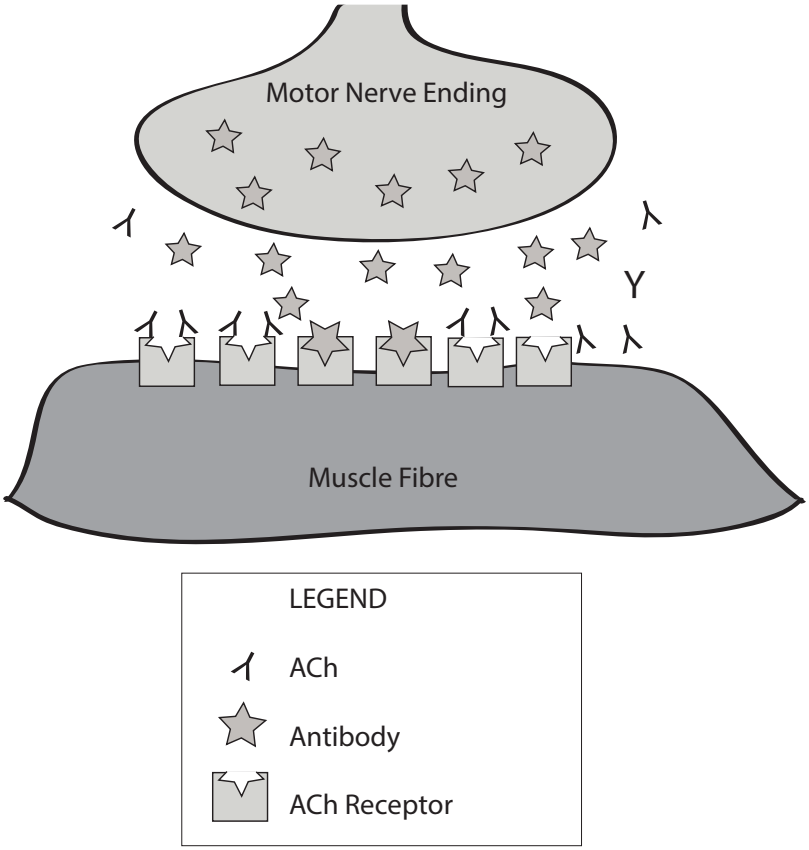
What causes Myasthenia Gravis?

Myasthenia Gravis is an autoimmune disease. The immune system normally makes antibodies (special proteins) to attack bacteria, viruses and other 'germs'. An autoimmune disease means that the immune system makes antibodies, which attack parts of the body. In the case of Myasthenia Gravis, these antibodies attack the receptors on the muscle fibres which means the muscles can't move properly. In most cases, patients with Myasthenia have antibodies to the acetylcholine receptor.

Who gets Myasthenia Gravis?

About 1 in 100,000 people in the UK develop Myasthenia Gravis each year. It can develop at any age.

Neuromuscular Junction



The Acetylcholine (ACh) is the chemical released by the nerve endings, which then latches onto the Acetylcholine receptor (ACh receptor) on the muscle surface.
The antibody is shown here as a star, attaching to the receptor in some cases.

What are the symptoms of Myasthenia Gravis?

The main symptom is weakness of muscles that can get worse with physical activity but improves with rest.

- Usually, the muscles around the eyes are affected first as they are used all the time and can tire quickly. This can lead to drooping of the eyelids and double vision.
- Muscles around the neck, face and throat are commonly affected. This can result in difficulty holding your head up. If the face and throat muscles are affected, there can be difficulty in swallowing, speaking and chewing.
- There may be weakness of the arms or legs.
- If Myasthenia Gravis is severe, it may affect your breathing muscles.

Complications

Sometimes, weakness of the breathing muscles can lead to a 'Myasthenic Crisis'. When this occurs, you will need to be admitted to hospital for treatment as this can be life threatening. The treatment may include being put on a ventilator to help your breathing.

Plasmapheresis is often used in the treatment of a Myasthenic Crisis. During Plasmapheresis, your plasma (plasma is the liquid part of your blood without the red or white blood cells) is exchanged for a blood donor's plasma, which doesn't have any abnormal antibodies. This can help for a short time.

An alternative to Plasmapheresis is Immunoglobulin treatment. This is a blood product, which is pooled from various blood donors, and helps to counteract your antibodies. The treatment is given via a drip, usually over 5

days. Both Plasmapheresis and Immunoglobulin treatment help to make your muscles stronger and rather quickly.

Types of Myasthenia Gravis

Myasthenia due to an antibody (immune-mediated)
About 15% of patients will only have eye muscle symptoms. We refer to them as **Ocular Myasthenia**.

Patients with weakness affecting muscles other than the eyes are referred to as having **Generalised Myasthenia**.

Most patients with Myasthenia have antibodies to the Acetylcholine receptor. However, a small percentage of patients with Myasthenia have a different antibody, which attacks Muscle Specific Tyrosine Kinase (or MuSK for short) instead of the acetylcholine receptor.

Some patients do not have detectable antibodies to the acetylcholine receptor or to MuSK. They are usually referred to as **Seronegative**. Although these patients still have an underlying antibody causing myasthenia, it is not found using the usual tests.

Thymus Abnormalities

The thymus gland is a small gland at the root of the neck just behind the upper sternum (breast bone). It forms part of the immune system. Many people with Myasthenia Gravis have an abnormal thymus, especially those who develop Myasthenia Gravis before the age of 40. It is thought that the thymus gland is the main source of the abnormal antibodies.

A small number of people with Myasthenia Gravis develop a tumour (growth) of the thymus gland called a thymoma. Usually, the tumour is benign (non-cancerous), but in a small number of cases it is malignant (cancerous).

Are tests needed?

- A blood test will help us confirm whether you have the abnormal antibody which causes Myasthenia Gravis
- In some cases you may need muscle and nerve tests to confirm a diagnosis
- A scan of the upper chest may be needed to look for thymoma (a tumour of the thymus gland). This occurs in a small number of cases.
- We sometimes check your blood to see if you have thyroid disease or other autoimmune diseases.

What are the treatments for Myasthenia Gravis?

Anticholinesterase Medicines Anticholinesterase medicines delay the breakdown of acetylcholine when it is released from the nerve endings. This means that more acetylcholine is available to fight against the abnormal antibodies, which attack the receptors on the muscle fibres. Anticholinesterase medicines work better when the disease is mild and the level of antibody in the blood is low, for example in people who just have eye muscle symptoms. The most common anticholinesterase medicine is called Pyridostigmine (Mestinon).

Thymectomy (removal of the thymus)
In some cases, removing the thymus is an option. This usually helps people under the age of 50, where the thymus gland produces the abnormal antibodies.

For people who:

- Are under the age of 50
- Have the antibody to the acetylcholine receptor
- Have widespread muscle symptoms

In half of the cases removing the thymus helps symptoms and for 3 in 10 people the symptoms are cured.
If the thymus appears enlarged on the scan, this could be a thymoma and nearly always should be removed.

Steroid Medication
Steroids suppress the immune system and stop the abnormal antibodies being made.

A low dose of steroids, usually every second day, helps if the symptoms are mild or if the disease only affects the muscles around the eyes. Higher doses may be needed if other muscles are affected.

It may take several months for the steroids to control symptoms. Once there is an improvement, the dose is slowly reduced to find the lowest dose needed to control symptoms. For some people, the dosage needed to control symptoms may be high which can lead to side effects.

Immunosuppressant Medicines Immunosuppressants work by suppressing the immune system and stopping the antibodies from being made. The most common immunosuppressant prescribed is Azathioprine, but others can be used.
You are often prescribed a steroid and an immunosuppressant as they work better together. This may mean you need a lower steroid dosage which in turn can mean fewer steroids.

What is the course of this treatment and outlook?

With Treatment
With treatment, most people can expect an improvement in their symptoms, with many people's symptoms 'disappearing'. In most cases, when the symptoms 'disappear', the treatment

such as steroids or immunosuppressive medicines can be stopped.

Life expectancy is normal.

Effects on Lifestyle

People with myasthenia Gravis are encouraged to lead a normal life and to try not to let it take over their life. They should eat healthy, but also be aware of any issues with their swallowing abilities which may require them to change the texture of their food. Alcohol should only be taken in moderation and smokers should try and stop as there are occasions when myasthenia gravis can affect the breathing muscles.

Further Help and Information

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