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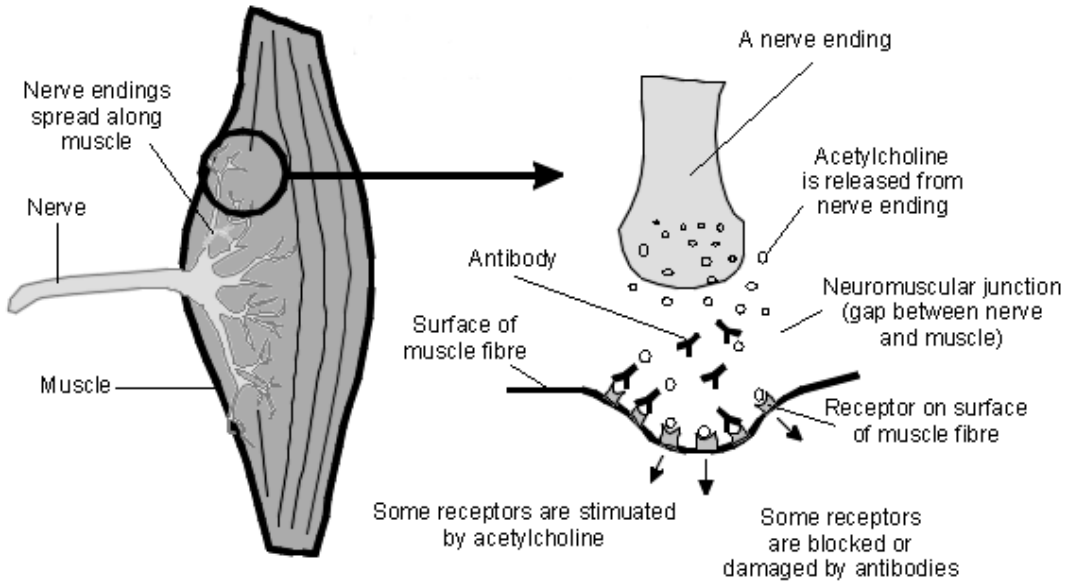
Myasthenia gravis

Myasthenia gravis (MG) is a condition where muscles become easily tired and weak. It is due to a problem with how the nerves stimulate the muscles to tighten (contract). The muscles around the eyes are commonly affected first. This causes drooping eyelids and double vision. Treatment is usually effective. However, relapses are common and many people with myasthenia gravis have long-term difficulties with their daily activities.

Understanding muscle

Each muscle is supplied by a nerve which splits into smaller nerves that spread along the muscle fibres. There is a tiny gap between the ends of the nerves and the surface of the muscle. This gap is called the neuromuscular junction (see diagram below).



The brain sends messages down the nerves to the muscles it wants to tighten (contract). The nerve endings release a chemical called a neurotransmitter into the neuromuscular junction. This neurotransmitter is called acetylcholine. The acetylcholine quickly attaches to receptors on the muscles. This in turn triggers the muscle to tighten. There are many acetylcholine receptors on each muscle fibre.

What is myasthenia gravis?

People with myasthenia gravis have a fault in the way nerve messages are passed from the nerves to the muscles. The muscles are not stimulated properly, so do not contract well and become easily tired and weak.

What causes myasthenia gravis?

Myasthenia gravis is an autoimmune disease. This means that the immune system (which normally protects the body from infections) mistakenly attacks itself. (Other autoimmune diseases include type 1 diabetes and thyroid disorders.) In most people with myasthenia gravis, antibodies are made which block, alter or destroy the acetylcholine receptors on muscles. The acetylcholine cannot then attach to the receptor and so the muscle is less able to tighten.

What causes the abnormal antibodies to be made?

The reason why the body's immune system starts to make abnormal antibodies against muscle receptors is not known. Abnormal antibodies are made in various parts of the immune system, including:

- The bone marrow.
- Blood.
- Lymph glands.
- The thymus gland.

However, the thymus gland is thought to be a main source of the abnormal antibodies (see below).

How common is myasthenia gravis?

Myasthenia gravis is a rare condition. It can occur at any age but most commonly affects women aged between 30 and 40 and men aged 50-70 years. Women are slightly more likely than men to have myasthenia gravis, particularly in certain ethnic backgrounds. It can occur in more than one member of the same family.

What role does the thymus gland have in myasthenia gravis?

The thymus gland is a small gland in the upper chest just behind the breastbone (sternum). It is part of the immune system. The thymus is abnormal in many people with myasthenia gravis, particularly in those who develop myasthenia gravis before the age of 40 years. The exact role of the cells in the thymus is not clear. However, the cells may have something to do with programming or making antibodies against acetylcholine receptors. For some people, surgical removal of the thymus gland by an operation cures their myasthenia gravis.

A small number of people with myasthenia gravis develop a growth (tumour) of the thymus gland, called a thymoma. If it occurs, it is usually non-cancerous (benign), However, in a very small number of cases it is cancerous (malignant).

What are the symptoms of myasthenia gravis?

The typical main symptom is weakness of muscles that becomes worse with activity and improves with rest. Affected muscles tire or become fatigued very easily. This means that symptoms are usually worse at the end of the day and after exercising. Symptoms may come and go at first when using the muscles. The symptoms vary enormously between people with myasthenia gravis:

- The muscles around the eyes are most commonly affected first, as these are constantly used and can quickly tire. This causes drooping of the eyelid (ptosis), and double vision. In some people, the muscles around the eyes are the only ones affected (when the level of abnormal antibody is low). If symptoms only affect the muscles around the eyes for longer than two years then the condition is unlikely to progress to other muscles. This is known as ocular myasthenia and affects 1 in 7 people with myasthenia gravis.
- Muscles around the face and throat are also often affected. Difficulty in swallowing and slurred speech may be the first signs of myasthenia gravis.
- Weakness in the arms, hands, fingers, legs and neck may develop.
- Weakness in the chest muscles sometimes occurs. If this is severe, a myasthenic crisis may result (see below).

The severity of symptoms (how easily the muscles tire) can vary from mild to severe. Infection or stress can make symptoms worse.

How is myasthenia gravis diagnosed?

It is not uncommon for the diagnosis of myasthenia gravis to be delayed. This is because muscle weakness often starts very gradually and can be mild at first. An examination by a doctor may find that you have muscle weakness which can suggest the diagnosis.

Tests that may then be done include the following:

- A blood test can detect the abnormal antibody and confirm the diagnosis in most cases.
- Muscle and nerve tests may be needed in some cases where the diagnosis is not clear. These usually happen in hospital settings.
- A scan of the upper chest may be performed to calculate the size and shape of the thymus gland.
- Breathing tests are performed in those people who have weakness of the chest muscles.

What is the treatment for myasthenia gravis?

In most cases, myasthenia gravis can be effectively treated.

Anticholinesterase medicines

These medicines delay the breakdown of acetylcholine when it is released from the nerve endings. More acetylcholine is then available to compete against the abnormal antibodies for the muscle receptors, which then improves the strength of the muscles. These medicines work best when the disease is mild and the level of antibody is low. The most commonly prescribed anticholinesterase medicine is called pyridostigmine. Diarrhoea is a common side-effect of this but it usually resolves without treatment.

Removal of the thymus (thymectomy)

This is an option in some cases. A thymectomy can improve symptoms for some people with myasthenia gravis.

Steroid medication

Steroid medication such as prednisolone tablets is often used in the treatment of myasthenia gravis. Steroids suppress the immune system and prevent the abnormal antibodies from being made. A low dose, often on alternate days, is usually enough for people where symptoms only affect muscles around the eye. Higher doses may be needed to prevent symptoms if muscles other than around the eyes are affected.

It may take several months to bring symptoms under control with steroids. Once improved, the dose is commonly reduced gradually to find the lowest dose needed to prevent symptoms. In some people, the dose of steroid needed to control the disease may be quite high and lead to side-effects. See the separate leaflet called Oral Steroids for more details.

Immunosuppressant medicines

An immunosuppressant medicine such as azothiaprine may be advised in addition to steroid medication. These medicines work by suppressing the immune system. If these medicines are not effective then a treatment called rituximab may be advised by your consultant.

Combinations of medicines

A steroid plus an immunosuppressant tends to work better than either alone. Also, the dose of steroid needed is often less if an immunosuppressant is added which reduces the risk of side-effects with steroids.

Myasthenic crisis

A myasthenic crisis occurs when the muscles that control breathing weaken to the point that breathing becomes very difficult. Admission to hospital is usually needed and sometimes assisted breathing by a ventilator machine is performed for a short while.

Plasma exchange may be given for a myasthenic crisis. This is a procedure where your plasma is exchanged for blood donor plasma which is free of abnormal antibody. This quickly gets rid of the abnormal antibody for a short while. This then gives time for other treatments described above to be started or modified.

What is the course of the disease and outlook (prognosis)?

The current available treatment options usually mean that people with myasthenia have a near-normal life expectancy. Without treatment, myasthenia gravis can become a serious, life-threatening disease. Treatment usually works well but many people with myasthenia continue to have some symptoms. Myasthenia gravis is a very variable condition and can cause long-term difficulties with daily activities.

Myasthenia gravis sometimes becomes less severe (goes into remission) temporarily, meaning the medication can be reduced or stopped. However, the symptoms often return (relapse).

Further reading

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Authored by:	Peer Reviewed by: Dr Toni Hazell	
Originally Published: 19/11/2023	Next review date: 29/09/2022	Document ID: doc_4586

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