South Devon Healthcare NHS
NHS Foundation Trust

PATIENT INFORMATION

Myasthenia Gravis

If you have any questions about the information in this leaflet please contact the

Neurology Department on
☎ (01803) 654827

What is myasthenia gravis?

Myasthenia gravis is a disease caused by antibodies which cause muscles to fatigue easily. Muscles are normally stimulated by a chemical called acetylcholine. This acts on receptors on the surface of muscles called acetylcholine receptors. The antibodies in myasthenia gravis bind to these receptors, competing with acetylcholine. The antibodies also damage the receptors in the long term. The effect is that muscles become weak and easily tired. These acetylcholine receptor antibodies are produced by an organ in the chest called the thymus gland. In some cases this gland is enlarged or cancerous. Occasionally other antibodies like anti MUSK antibodies may cause myasthenia gravis.

What are the symptoms of myasthenia gravis?

Myasthenia gravis may affect just the muscles around the eyes (ocular myasthenia). This results in droopy eyelids and double vision. When other muscles are also affected (generalised myasthenia) other symptoms develop. These include difficulty swallowing, slurring of speech, limb weakness, difficulty chewing, difficulty keeping the head up, jaw dropping, and difficulty with breathing. Ocular myasthenia may progress to become generalised.

The weakness in myasthenia gravis typically fluctuates, and is worse after the muscles have been in use, and better after resting. Symptoms are therefore worse when tired and later in the day.

How is myasthenia gravis diagnosed?

The history of fatigable muscle weakness is typical of myasthenia gravis. This is confirmed by blood tests looking for the presence of antibodies which cause myasthenia gravis. These antibodies are however not detectable in all cases. Electrical studies of the muscles called EMG may show characteristic abnormalities in myasthenia gravis. A CT scan of the chest is done to look for enlargement or cancer of the thymus gland. This may be repeated about 5 years later if it is normal the first time around.

How is myasthenia gravis treated?

Immediate treatment aims to increase the amount of acetylcholine to overcome the action of the antibodies. This is done with a drug called Pyridostigmine. This prevents the breakdown of acetylcholine and needs to be taken three to four times a day.

Other treatments reduce the production of the antibodies that cause myasthenia gravis by suppressing the immune system. These drugs include Steroids which are used for a short time because of side effects when used for prolonged periods. Other immune suppressing medicines include Azathioprine and Methotrexate; these are used for longer periods.

In severe cases of myasthenia gravis where the antibody levels are very high, treatment is given to mop up the antibodies. This can be done by an infusion of Immunoglobulins, which are blood products, or by Plasma Exchange which cleans the blood of the antibodies. Surgery to remove the thymus gland may also be effective even if the thymus is not obviously abnormal.

What does a patient with myasthenia gravis need to do?

If you have myasthenia gravis, your Neurologist will prescribe one or more of the above treatments. You should comply with these to prevent worsening or generalisation of symptoms. You should have your regular seasonal flu vaccinations.

You should be careful to avoid some medications that can worsen myasthenia gravis. Some of these medicines are listed at the back of this leaflet. Some anaesthetic medications used in surgery may also worsen myasthenia and your surgeon and anaesthetist need to be aware of your diagnosis if surgery is planned for any reason.

You should carry an alert card or bracelet to let people know you have myasthenia gravis in case of emergencies when you are unable to communicate. You also need to inform the DVLA of the diagnosis.

What do patients on pyridostigmine need to watch out for?

Pyridostigmine may cause stomach cramps and your Neurologist may prescribe a medicine called Propantheline to counteract this. Pyridostigmine may also cause diarrhoea and rarely muscle twitching. Excessive dosing with pyridostigmine may also cause your weakness to get worse.

What do patients on Steroids need to do?

Steroids may thin the bones (osteoporosis) and your doctor will prescribe Vitamins D, Calcium and medications called Bisphosphonates to protect and strengthen the bones.

Steroids may also irritate the stomach and your doctor will also prescribe gut protecting medications, usually Omeprazole.

If you need to take steroids for long periods, you will be referred for regular bone scans, usually every year, to monitor for bone thinning. You can also protect your bones by avoiding smoking and excessive alcohol. Steroids predispose to diabetes, high blood pressure and weight gain, and these would be monitored regularly by your doctor.

What do patients on immune suppressing drugs need to do?

Immunosuppressants like Azathioprine and Methotrexate can affect the function of the liver, kidneys and bone marrow. Regular blood tests are required before and during treatment with these medications. There is an increased risk of infections on immune suppressing drugs and, in addition to the seasonal flu vaccinations, a 5 yearly pneumococcal vaccination is recommended; this can be obtained at your GP surgery. You should avoid live vaccinations as these may result in infection. Live vaccines include BCG, varicella, measles, mumps, rubella, oral typhoid and oral polio.

What do patients on Azathioprine need to do?

There is a small risk of skin cancer with Azathioprine and this is linked to excessive sun exposure. If you are on Azathioprine you should avoid direct sun exposure, and use sunscreens and head protection when appropriate. You should report any new skin changes to your doctor. Azathioprine may also harm the foetus of pregnant women; if you are at risk of getting pregnant, you should be on contraceptives to prevent this.

What do patients on Methotrexate need to do?

Methotrexate may cause lung damage. To make sure that your lungs are not already damaged your Neurologist will arrange a chest X-ray before you start Methotrexate, unless you have had one within the previous 6 months. Methotrexate may also harm the foetus and contraception is advised if there is a chance of pregnancy. Methotrexate may also damage sperm cells in men; to prevent this resulting in malformed babies, male or female contraception should be used during, and for at least three months after the drug is stopped.

Medications to avoid or use cautiously in myasthenia gravis

- Aminoglycoside antibiotics like Gentamicin
- Other antibiotics like Erythromycin and Ampicillin,
- Botulinum toxin
- Beta blockers like propranolol
- Heart rhythm medicines like Quinidine and Procainamide
- Trimethaphan
- Lidocaine
- Phenytoin
- Chloroquine
- Lithium
- Magnesium
- Oxytocin
- Diazepam
- Penicillanine
- Ketamine

Where can I get more information and support on myasthenia gravis?

The Myasthenia Gravis Association

(www.mga-charity.org)

The South West Neuromuscular Network
(www.swscg.org.uk/networks/Neuromuscular/Neuromuscular.htm)

Myasthenia gravis management checklist

Tests

- ☐ Acetylcholine receptor antibodies
- ☐ EMG
- ☐ CT chest; if initially normal consider repeating in 5 years
- ☐ MPTP before azathioprine
- ☐ Hepatitis screen before Azathioprine
- ☐ Baseline Bone Mass Density screen and annually
- ☐ Chest X ray before Methotrexate

Advice to patients

- ☐ Patient information leaflet
- ☐ Contraceptive advice on immunosuppressants
- ☐ Sun protection on azathioprine
- ☐ Avoid allopurinol on azathioprine
- ☐ Annual flu
- ☐ 5 yearly Pneumovac if on immunosuppressants
- ☐ DVLA advice
- ☐ Advice on drugs to avoid

Steroids

- ☐ Explain steroid risks
- ☐ Start steroids at 10-25mg daily and keep alternate daily
- ☐ Daily steroids if with diabetes
- ☐ Bone and gut protection

Monitoring

- ☐ Shared care agreement form
- ☐ Consider monitoring antibody when cutting down treatment
- ☐ Monitor BP and weight on steroids
- ☐ B12, Folate and TSH if MCV >105
- ☐ Consider Thymectomy if no response to pyridostigmine

References

1. Skeie GO, Apostolski S, Evoli A, et al. Guidelines for treatment of autoimmune neuromuscular transmission disorders. Eur J Neurol 2010; 17: 893-902.
2. Chakravarty et al. BSR/BHPR guideline for disease modifying anti-rheumatic drug (DMARD) therapy in consultation with the British Association of Dermatologists. Rheumatology 2008; 1-16.
3. Meggitt et al. British Association of Dermatologists guidelines for safe and effective prescribing of azathioprine 2011. Br J Dermatol 2011; 165: 711-734.