

Myasthenia Gravis

A comprehensive guide to living with and understanding
this manageable condition.



Proudly compiled and provided by the
Myasthenia Gravis Association of Queensland Inc.

ABOUT THIS GUIDE

As each person's experience with Myasthenia Gravis (MG) is unique, this guide can only approach the topic in a general way. Many patients want to be fully informed about the nature of this condition and this booklet provides information and guidance, not only to those suffering from MG, but also to their families, friends and anyone interested in finding out more about the condition.

This booklet deals in detail with Myasthenia Gravis and mentions Ocular Myasthenia, Lambert Eaton Myasthenic Syndrome and Congenital Myasthenia.

Whilst this handbook provides important and helpful information it is not intended to replace professional advice. Diagnosis and advice on medical care and other assessments should be sought from the appropriate medical professional/s. Each case is different and only the treating professional can advise in individual situations.

As a patient of MG, a relative or a carer, you may find the science of MG and associated medical terms unfamiliar. A 'Glossary of Terms' has been included.

The Myasthenia Gravis Association of Queensland Inc. was formed in 1991 and has been recognised by Queensland Health as the peak body for Myasthenics in Queensland. MGAQ is an incorporated association (IA100400), a registered charity (CH1212) and our ABN is 92 055 613 137. More information is available on our website www.mgaq.org.au.

The Myasthenia Gravis Association of Queensland Incorporated has approved this publication for supply, free of charge, to sufferers of myasthenia gravis, medical practitioners and other professionals who look after them. Copies can be obtained from the association by phoning **1800 802 568**, emailing info@mgaq.org.au or visiting the website at www.mgaq.org.au



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PART ONE

MYASTHENIA GRAVIS - AN INTRODUCTION

What is Myasthenia Gravis?

(Definition courtesy of the National Institute of Neurological Disorders and Stroke USA)

Myasthenia Gravis is a chronic autoimmune disease characterised by varying degrees of weakness of the skeletal (voluntary) muscles of the body. The name 'myasthenia gravis', which is Latin and Greek in origin, literally means "grave muscle weakness". With current therapies, however, most cases of myasthenia gravis are not as 'grave' as the name implies. In fact, for the majority of individuals with myasthenia gravis, life expectancy is not lessened by the disorder.

The hallmark of myasthenia gravis is muscle weakness that increases during periods of activity and improves after periods of rest. Certain muscles such as those that control eye and eyelid movement, facial expression, chewing, talking and swallowing are often, but not always, involved in the disorder. The muscles that control breathing and neck and limb movements may also be affected.

Myasthenia Gravis occurs in all ethnic groups and both genders. It most commonly affects young adult women (under 40) and older men (over 60), but it can occur at any age.

When Was Myasthenia Gravis First Diagnosed?

Myasthenia Gravis (MG) was first clearly described in the 1600's by Dr Thomas Willis, a well-known London physician, who wrote the following:

"...in the mornings, they are able to walk firmly...Or to take up any heavy thing. Before noon, the stock of the spirits being spent, which had flowed into the muscles, they are scarce able to move hand or foot.... A prudent and honest woman has this spurious palsie (sic) since many years, not only in her members (limbs)....But, after she has spoke long, hastily or eagerly, she becomes as mute as a fish; nor can she recover the use of her voice under an hour or two..."

From that time until the 1800's progress on finding out more about this condition was slow. In Europe, neurology became a separate specialty in about 1860 and myasthenia gravis was first recognised in its own right about 1880 by German Neurologists Wilhelm Erb and Friedrich Jolly. These men made the distinction between Motor Neurone Disease (MND) and Myasthenia Gravis (MG). Unlike MND, MG was not remorselessly progressive, but rather was fatiguable – the harder one tries, the weaker one becomes. It was first called 'Myasthenia Gravis' in about 1895.

Since then, the discoveries progressed along two parallel paths and then came together only in the 1970's.

- A. The first was the study of how nerves trigger muscles. In the 1850's in France, Claude Bernard showed that an arrow poison 'Curare' worked by blocking this triggering of the muscles.

In the 1860's, Dr Fraser and his team in Edinburgh developed an antidote for Curare using the Calabar Bean poison. This paved the way for the production of purified Physostigmine. The modern Neuromuscular Blocking Agents are derivatives of curare.

Then, in 1934, Dr Mary Walker noticed that patients who had myasthenia gravis looked like patients who had been given too much curare. She used this information to show that physostigmine helped to strengthen muscles. She was involved in the development of pyridostigmine (ie Mestinon®) which came into use in 1954. Dr Mary Walker's breakthrough also helped to explain the underlying defect - it proved that MG results from a defect in nerve/muscle triggering.

- B. The second starting point was the thymus, an organ behind the breast bone whose function was then a mystery. Thymic tumours (thymomas) were first noticed in some myasthenics in 1899 by H. Oppenheim in Berlin and C. Weigert in Frankfurt. At that time, it was too dangerous to operate on the chest because there was no way to keep the patient breathing when the rib cage was opened. With the development of positive pressure ventilation an American surgeon, Alfred Blalock, in 1939, was able to remove a thymoma from a patient whose MG improved dramatically afterwards. It was later found this marked improvement so soon after surgery was unusual and improvement is usually seen after an extended period of time. Over the next twenty years a very distinguished surgeon, Sir Geoffrey Keynes, documented his first 155 thymectomies and noted that improvements were mostly confined to myasthenics without thymomas.

Finally, in the 1960's Dr Jacques Miller (London and Australia) and Dr Bob Good (USA) showed that the thymus was the key 'immune

organ'. It generates 'T cells' that help to switch on other immune cells to make antibodies and/or destroy germs. In 1959, Professor Iain Simpson, in reviewing the medical history of people who had thymectomies, noticed that such autoimmune diseases were especially common in relatives of myasthenics. So he proposed, in 1960, that MG was an autoimmune disease also.

The above two paths finally came together in 1973. By then medical experts had realised that the ignition keys (Ach) must somehow latch onto specialised Ach ignition locks (the AChR receptors). Scientists experimented with snake venom to purify the AChR receptors in electric fish. From this experimentation they were able to prove conclusively that myasthenia gravis could be caused by antibodies.

This understanding of the role of antibodies led to the use of plasma exchange (plasmapheresis) to wash the antibodies out of a patient's bloodstream. About four litres of the patient's blood is drained a litre at a time and placed in a centrifuge where the plasma, in which the antibodies are found, is removed. The plasma is replaced by fresh plasma, thus reducing the antibody level.

This breakthrough in understanding the role of antibodies also led to the development of immune-suppressive drugs such as Azathioprine. It also provided a basis for treating myasthenia gravis with steroids. They were already in use for MG and, by the 1960's, it became clear that they reliably lower the antibody levels within about three months.

A recent observation has been the increased incidence of older people, especially women, being diagnosed with myasthenia gravis. No definitive reason has yet been advanced except that people are living longer. Not all women, when tested, prove antibody positive and, in most cases, the thymus gland is no longer active. However, it has been established that, in a number of these patients, a protein presence has been found at the neuromuscular junctions. This protein is called Muscle Specific Tyrosine Kinase (MuSK).

There is still a lot about myasthenia gravis that is not understood, especially about how an immune attack starts and how it can be turned off selectively in order to not damage all the protective immune responses.

What are the Causes of Myasthenia Gravis?

In order to make a muscle contract, the brain sends an electrical signal through the nervous system to the nerve endings that lie next to the muscle fibres that make up the muscle. There is a tiny gap between the nerve endings and the surface of the muscle fibres.

The electrical signal from the brain triggers the release of a chemical (acetylcholine or ACh) at the nerve endings. The acetylcholine crosses the gap between the nerve endings and muscle fibres and attaches itself to special receptor cells on the fibres, thus making the muscle contract. Another chemical (acetylcholinesterase or AChE) then breaks down the acetylcholine, making the muscle relax.

Myasthenia Gravis is an autoimmune disease because it is caused by the body's own immune system which attacks itself. If a person has MG, the body's immune system produces antibodies that block or damage the muscle receptor cells, reducing the number available on the muscle fibres. When this happens, the normal communication between the nerves and muscles are affected, the muscles do not contract well and they become weak and easily tired.

The reason that some people's immune system makes antibodies that act against muscle receptor cells is not fully understood. It is thought that the thymus gland – part of the immune system located in the upper chest – may be linked to the production of these antibodies. Approximately 10% of people with myasthenia gravis have a benign tumour of the thymus gland.

Myasthenia Gravis can be triggered in some people by particular viruses or medicines. It is also thought that some people's genes make them more likely to develop autoimmune diseases.

The myasthenias come in quite separate forms:

- **Myasthenia Gravis (MG)** - is by far the commonest form of myasthenia. Here, an immune attack damages the muscle receptor cells of our voluntary muscles only. The weakness typically fluctuates and is 'fatiguable' – the more you try, the worse it gets. Thus, patients are often stronger in the mornings and get weaker during the day.
- **Ocular Myasthenia** - where the condition is only seen in the eye muscles. Myasthenia Gravis can affect one group of muscles much more than others, for example just one small muscle that moves one eye in one direction. This weakness may be the only problem in some patients or could be an early sign of further muscle weaknesses in others. Strictly speaking, the label of 'Ocular Myasthenia' is only given if the weakness is still restricted to the eye movements at least two years after the first symptom.

- **Congenital Myasthenia (CM)** - accounts for less than one in twenty of all myasthenias. Here, inherited faults make the ignition system less efficient. People are born with this condition and the onset of symptoms may appear shortly after birth or may not show for some years. CM is a faulty gene affecting the nerve-to-muscle signalling and these myasthenias do run in families. As such, faults occur at random in any gene, so they can affect any system in the body, and vary greatly. Because this is not an autoimmune problem, none of the immune treatments is suitable. However, non-immunological treatments such as Mestinon® may still be useful in alleviating weakness.
- **Lambert-Eaton Myasthenic Syndrome (LEMS)** - accounts for approximately one in twenty of all myasthenics. LEMS is rare and usually starts after age 30 years. Here, a similar immune attack damages the nerve endings in both the voluntary and the 'automatic' muscles (eg in the blood vessels, bowel and bladder). It also affects the limbs more than the head, neck and trunk. This condition causes weakness at the neuromuscular junction and, unlike MG, some people with LEMS improve with exercise. LEMS is often a paraneoplastic condition, ie. may be associated with a tumour, especially lung.
- **Neonatal Myasthenia** - in some cases, babies born to myasthenic mothers may, for a brief period of about four weeks, exhibit transient myasthenic symptoms. They fully recover if properly supported during this period while the mother's antibodies clear from the baby's bloodstream.

None of the above myasthenias directly affect sensation (eg sense of touch or temperature) or cause pain (though overstrain may cause aches in areas such as the back and neck).

What are the Symptoms of Myasthenia Gravis?

No two patients show exactly the same symptoms, either in kind or in severity. The onset can be sudden but, much more commonly, it starts so gradually and insidiously that it is missed or diagnosed only after a period of time. For most people with myasthenia gravis, symptoms are mild at first but get steadily worse over several months, reaching their most severe within the first two years and then levelling off.

The muscles which we use all the time such as those which keep our eyelids open are often, but not always, the first to indicate that something is wrong. Troubles with facial muscles, the ones used to smile, speak and swallow are also among the first to signal that there is a problem. Other symptoms that may, or may not, be present include:

- **Blurred or double vision;**
- **Slurred or nasal speech;**
- **Weak or droopy eyelids;**
- **Weakness of the facial muscles causing a ‘snarling’ smile;**
- **Difficulty breathing, particularly when exercising or lying flat;**
- **Difficulty with neck leading to difficulty holding up the head;**
- **Tiring easily just from the act of chewing and swallowing;**
- **Difficulty swallowing food and/or drinks;**
- **Unstable gait;**
- **Balance problems;**
- **Weak or fatiguing hand and arms leading to difficulty lifting, etc;**
- **Weakness in the legs leading to difficulty walking;**
- **General fatigue or fatigue brought on by physical exercise.**

As myasthenia gravis muscle weakness becomes worse when the affected muscle/s is used and improves when it is rested, symptoms often get worse as the day goes on but may get better with rest or a good night's sleep. Stress, hot weather and infection can also make muscle weakness worse and women may find that their symptoms are more severe during their period. Also, MG has a ‘head downwards’ bias so the nearer the feet, the milder it is and the less likely muscles are to be affected.

Myasthenia Gravis is not usually painful although, if it affects the neck muscles, the back of the neck can become painful from the effort of holding up the head.

Occasionally, muscle weakness can cause severe swallowing or breathing problems (known as a myasthenic crisis) and requires urgent medical attention.

Who Gets Myasthenia Gravis?

Myasthenia Gravis presents at any age. Female incidence peaks in the third decade of life whereas male incidence peaks in the sixth or seventh decade. The mean age of incidence of MG is 28 years in females and 42 years in males. As patients often have the condition for many years and grow older with it, there is actually more older women than younger women with the condition at any time.

Myasthenia Gravis is not contagious, nor is the acquired form with AChR antibodies considered to be hereditary. It is known, however, that other siblings in a family have an increased risk of being diagnosed with MG. Given the low incidence of the disease the chances of siblings experiencing MG are still uncommon, occurring in approximately 1% of cases.

One of the inherited risk factors for 'early-onset myasthenia gravis' (starting before age 40, mostly female) also predisposes the patient to other autoimmune disorders, so thyroid disease and 'young-onset' diabetes are slightly more common in myasthenic patients – and also their blood relatives – than in the national average. Other autoimmune diseases that occur in higher frequency in patients with MG are hyperthyroidism, rheumatoid arthritis, scleroderma and lupus.

Other inherited and external risk factors such as infections are suspected (but unproven) to also contribute to the onset of myasthenia gravis but very little is known about the why and how. Internal factors include thymic tumours ('thymomas') in approximately one myasthenic patient in ten, and thymic changes in 'young-onset' patients.

PART TWO

MYASTHENIA GRAVIS – DIAGNOSIS AND TREATMENT

How is Myasthenia Gravis Diagnosed?

Myasthenia Gravis affects people initially in a variety of ways so that no two cases have exactly the same symptoms. The onset can be sudden or, more commonly, it starts gradually and develops slowly or intermittently. This, plus the fact that its symptoms can be confused with other ailments, makes it easily missed or only diagnosed after some time.

Positive diagnosis is now available, even though this is normally only achieved over a period of time and, in some cases, requires confirmation by more than one test.

The patient's account of their symptoms, and the visible pattern of weakness are tell-tale signs of suspected myasthenia gravis. Once suspected it can be confirmed in several ways:

- **Fatigue** – observation of repetitive movements of the eyes, arms or legs. This can be done without equipment.
- **EMG (Electromyography) and SFEMG (Single Fibre Electromyography)** – fatigue can also be measured electrically by recording the responses of a muscle to stimulation of its nerve with harmless electrical needles – electromyography. 'Single Fibre EMG' provides the most sensitive test. A tiny needle is placed with a number of individual muscle units (of which there are hundreds or thousands in each muscle) and the firing of each muscle unit is observed. EMG also helps to avoid confusion with LEMS and Congenital Myasthenias which may require different treatments.
- **Ice Pack Test** - cooling may improve neuromuscular transmission. In a patient with myasthenia gravis who has ptosis (droopy eyelid), placing ice over an eyelid will lead to cooling of the lid, which leads to improvement of the ptosis.
- **Tensilon Test** – Edrophonium®, a cousin of Mestinon®, is a short-acting anti-cholinesterase drug that is injected into the worst affected muscle groups (usually two of them). Strength is measured before and after the injection. This Tensilon test is used less often nowadays as it carries some risks in the very rare case where someone is allergic to edrophonium (this drug can potentially cause an allergic reaction or anaphylaxis). Thus, the test is best done in a hospital with equipment ready in case of emergency. Alternatively, the patient's

general improvement on anti-cholinesterase drug treatment is useful supporting evidence.

- **Blood Test** – the most specific diagnostic test is a blood test for the typical antibodies to the AChR. Approximately 85% of all patients with myasthenia gravis have these antibodies whereas patients with other muscle diseases almost never do. It has been recently discovered that about half of these patients who are negative to AChR have antibodies to another protein present at the neuromuscular junction called Muscle Specific Tyrosine Kinase (MuSK). The role of the MuSK protein is still being clarified but it is known that it plays a critical role in postsynaptic differentiation and clustering of acetylcholine (ACh) receptors. Patients with anti-MuSK antibodies are predominantly female.
- **CT Scan or MRI** - of the chest is highly accurate in detecting thymoma. Every patient with myasthenia gravis should be screened for thymoma. Chest radiography is relatively insensitive in screening for thymoma, as it does not detect up to 30% of cases.

What Does it Mean to Have Myasthenia Gravis?

In the past (pre 1960), untreated myasthenia gravis carried a mortality rate of 30 -70%. In the modern era, patients with MG have a near-normal life expectancy.

Generally, myasthenia gravis is a persistent condition requiring chronic treatment. Fluctuations over the long term are the norm. The neuromuscular junction cannot be re-formed, unlike many parts of the nervous system. Muscle strength that has been affected by MG for a long time often recovers with treatment. This means that the intensity of treatment for MG can be modulated to the current severity of the condition.

Now, with greatly improved treatment, myasthenia gravis rarely shortens life and most patients can lead an active life, despite a few side-effects from the treatments. Unfortunately, only approximately one patient in five goes into remission and most have to learn how best to manage it.

How is Myasthenia Gravis Treated?

During the past seventy years, myasthenia gravis has gone from being an almost unknown fatal disease to the current situation in which the symptoms can be controlled so that we have ageing myasthenics in the community.

Treatment is aimed at relieving symptoms. The patient's medical specialist carefully measures strength in various muscles and muscle groups to determine severity and extent of the disease and to monitor the benefits of treatment.

There is a range of treatments available for treating myasthenia gravis, although often they are used together:

- **Pyridostigmine (Mestinon®)** – this is usually the first-line treatment for MG. It is a reversible inhibitor of acetylcholinesterase (AChE) so increases acetylcholine (ACh) stimulation of the remaining acetylcholine receptors. If there is insufficient acetylcholine receptors remaining to trigger the muscle action, the Mestinon® is not going to help. The use of Mestinon® does nothing to cure MG or attack the rogue antibodies, but assists the patient in coping by improving muscle strength temporarily with each dose. Other drugs in the same category are ephedrine and spironolactone.

Mestinon® comes in 60mg and 10mg tablets, as well as a slow release 180mg dose which is usually used for nocturnal symptoms. Mestinon® assists the patient to a level where optimum strength is restored, but attempts to raise that level of strength by increasing the dose may have the reverse effect and actually increase the weakness. If the dose is too high a Cholinergic Crisis may occur (this almost never happens to people taking fewer than six 60mg tablets per day). Obviously, it is important to distinguish that from a myasthenic crisis, which needs completely different treatment (eg more Mestinon®). The dosage for one person may be very different from that required for another person and a degree of trial and error and working with the relevant medical specialist may be required in order to arrive at the best result. Since Mestinon's® effects last only a few hours, it is important that it be taken as directed and patients should always carry some medication with them. The taking of Mestinon® can sometimes cause stomach cramps and diarrhoea so taking it with bland food or 30 minutes in advance of eating food may help. If ongoing stomach cramps, diarrhoea, flickering eye movements or body cramps are experienced, the patient should consult with their medical practitioner.

- **Immunosuppressants** – the principal drugs used to suppress the immune system in myasthenia gravis are a steroid such as prednisone and an immune-suppressant such as azathioprine. The response to these treatments can take weeks to many months, with the maximum effect taking months to years. By suppressing immunity generally, the increased risk of infection must be recognised.
 - **Corticosteroids (Prednisolone, Prednisone, Hydrocortisone, Dexamethasone)** - are known to reliably improve myasthenia gravis after a delay of two to six weeks. Prednisolone is a synthetic hormone commonly referred to as a 'steroid' and is very similar to the hormone 'cortisone' which is produced naturally in the body. Steroids suppress the production of antibodies. This suppression can make it slightly harder for a person to fight off infection, but also stabilizes the immune system if it is overactive. It is generally known that, with chronic use, these drugs have side effects. Once again, the determination of a maintenance level is only achieved through trial and error with a medical professional. Severity of MG, control of the symptoms and the development of adverse effects are all taken into consideration when the medical specialist determines the dosage of prednisolone. Deterioration in MG can occur in the first few weeks of treatment so the dose is often increased slowly and with ongoing surveillance.
 - **Azathioprine** - this drug also reduces antibody production, but it takes much longer to take effect (at least one year). It is sometimes used by itself with patients who cannot manage on Mestinon® alone. More often, it is used to enhance the benefits of steroids in order to manage with lower steroid doses. Some patients cannot take azathioprine due to rash, hepatitis, nausea or vomiting but this is usually evident within two weeks to two months of commencing the medication.
 - **Cyclosporine** - this drug suppresses the autoimmune response that is responsible for causing fluctuating and fatigable muscle weakness. Some MG patients may notice a gradual improvement in their symptoms after a period of 2-3 months. Others take longer to see a response. Cyclosporine is manufactured in both a capsule and oral form.
 - **Other immunosuppressant drugs** including **methotrexate**, **cyclophosphamide** and **mycophenolate** may also be used to treat MG and are usually given in conjunction with steroids.

- Plasmapheresis (Plasma Exchange)** – is a procedure where blood is separated into cells and plasma (the liquid which contains the antibodies). The plasma is removed and replaced with a blood product called albumex which is made from human albumin, or fresh frozen plasma if needed. This treatment is used to stabilise rapid decreases in muscle strength or to add to present treatment if current forms of therapy are providing insufficient control of the disease. It is also used to reduce moderate to severe muscle weakness before surgery. The number of plasmapheresis treatments needed depends on the protocol the physician has determined is best for the patient. For plasmapheresis to occur a needle is placed in each arm. If the arm veins are too small to use, and the treatment period likely to be prolonged, the physician may insert a portacath or create a fistula. Unless the physician has instructed otherwise, it is important to eat before the plasma exchange and not skip any meals. During plasma exchange, the patient may drink fluids. The patient should empty the bladder prior to the procedure. Wearing comfortable clothing with loose fitting sleeves that pull easily above the elbows will make it easier to place the needles in each arm. Taking something to read will help pass the time and, because the albumex may be cold when inserted into the veins, socks and a light cardigan are suggested. The time spent on the machine may be one to three hours, depending on the patient's weight, height and the amount of plasma to be exchanged. Most patients feel fine after the procedure but, if not staying in hospital, someone should be available to drive the patient home. Plasmapheresis works quickly to increase strength and most patients begin to improve after the first few days of the treatment. Some common side effects include a high temperature, a drop in blood pressure, tingling associated with the mouth, eyes, fingers or toes and a possible allergic reaction to the solution which may result in itching, wheezing or rash. Plasmapheresis must be done under close medical supervision. As with all medical procedures, blood component therapy involves some risks. These are limited as far as possible through the care that is taken when the blood is collected and processed. All blood donors are checked for viruses, people who have been in contact with hazards to the blood supply are not allowed to donate blood, and surveillance systems are in place to detect and respond to other risks if they emerge. Statistics show that the likelihood of infection from a blood transfusion is the same as the likelihood of being killed in a road accident. Also, Australia has one of the safest blood supplies in the world in terms of viral safety.
- Intravenous Immunoglobulin (IVIg)** – is also known as *pooled human gamma globulin* or simply *gamma globulin*. Immunoglobulin is a purified blood product made by 'pooling' the antibodies (Ig) from a large number of healthy blood donors and then slowly injecting it into the vein (IV). IVIg seems to affect the function or production

of antibodies in the immune system. The exact mechanism of how IVIg works in successfully treating myasthenia gravis and other autoimmune disorders is not entirely understood. Like plasmapheresis, it needs to be repeated at least every few weeks and is very expensive to produce as it relies on many, many blood donors. However, unlike plasmapheresis, it only requires a small intravenous line to be given. Wearing comfortable clothing with loose fitting sleeves that pull easily above the elbow will make it easier to place the needle in the arm. Taking something to read will help pass the time and, as most hospitals are air-conditioned, socks and a light cardigan may be warranted. The time spent on the machine is usually about three hours but may be longer depending on the patient's weight. As with plasmapheresis, IVIg is felt to be very safe with regard to exposure to infection or viruses. Blood donors are screened and the processing of IVIg inactivates such infections as HIV, Hepatitis B and C. Nonetheless, it is a human blood product that comes from multiple donors. Patients sometimes get a headache, a rash or rise in blood pressure. Blood tests are used to monitor for possible effects on the kidneys.

- **Thymectomy** – myasthenia gravis is often associated with enlargement of the thymus gland and this gland is seen as the producer of the rogue antibodies. Normally, the thymus helps the body's immunity by recognising and destroying any 'intruders' from a splinter in the finger to bacteria, viruses, cancer cells etc. A thymectomy is performed almost always for thymic tumours (thymomas) to prevent them from spreading and is also sometimes performed on myasthenics without significant thymoma. The thymus gland is located in the upper chest under the breastbone. It is composed of two main lobes and is shaped like a butterfly over the windpipe. The exact role of the thymus in MG is not completely understood. The surgery can be performed in several ways and the surgeon will determine the optimal procedure which may be based on position or number of lobes of the thymus. Convalescence after surgery is usually four to six weeks.

What are the Possible Side Effects of Some Drugs Used in the Treatment of Myasthenia Gravis?

Most medicines and drugs have some unwanted side effects. This includes those used to treat myasthenia gravis where the side effects will vary from patient to patient and with the level and frequency of medication. However, generally the benefits of the treatment outweigh the risks. Any adverse reactions should be reported to the MG-treating physician.

Dentists, optometrists, podiatrists and any health professionals with whom the patient has contact should also be aware that they suffer from myasthenia gravis and the medication they have been prescribed.

Some side effects to be aware of are:

- **Mestinon® (pyridostigmine) and Prostigmin® (neostigmine)** – not all Mestinon® preparations are equal. Mestinon Timespan® should never be substituted for regular Mestinon®. No single fixed dose schedule will suit all patients with myasthenia gravis whose medication requirements vary from time to time, day to day and in response to stress or infection.

Mestinon® is a reversible inhibitor of acetylcholinesterase (AChR) so increases acetylcholine (ACh) stimulation of the remaining acetylcholine receptors. ACh also plays an important role in many parts of the body in addition to the muscle receptors. This medication is not selective and thus can cause a number of unwanted effects including:

- **Gastrointestinal upset;**
 - **Nausea, vomiting;**
 - **Abdominal cramps and diarrhoea;**
 - **Increased salivation and tearing;**
 - **Increased bronchial secretions;**
 - **Increased sweating;**
 - **Muscle cramps;**
 - **Muscle fasciculations (twitching);**
 - **Muscle weakness;**
 - **Headache.**
- **Imuran® (azathioprine)** – this drug suppresses the immune system thus causing a reduction of the antibodies responsible for myasthenia gravis. The drug is extremely useful as it allows smaller doses of other drugs, particularly steroids. Unfortunately, as well as reducing the antibodies, Imuran® also reduces the formation of new blood cells. This effect must be monitored by regular blood tests. Liver function can also be affected by Imuran® but the damage is reversible on stopping the treatment or reducing the dosage. Because long term use may be associated with an increased risk of skin cancers, patients should also take extra care with their skin when outdoors and avoid going out in the sun as much as possible. Regular (eg yearly or more frequently in high risk cases) skin inspection with your GP or dermatologist is advisable.

Imuran® is tolerated by most people but patients should contact their treating specialist should any of the following occur:

- **Nausea and vomiting;**
 - **Fever and chills (flu-like symptoms);**
 - **Cough or shortness of breath;**
 - **Upset stomach, including diarrhoea;**
 - **Skin rash;**
 - **Darkening of the skin;**
 - **Cold sores in the mouth;**
 - **Blood in the urine or stool;**
 - **Yellowing of the skin and eyes.**
- **Steroids** – Prednisolone and prednisone should not be confused with anabolic steroids which are prohibited for use by athletes etc. Those used in medicine are usually much stronger than those occurring naturally in the body. Adverse effects from the use of steroids may include:
 - **Insomnia and mood changes** - euphoria or depression may occur and the cause is not clearly understood. It is best to take steroid medication in the morning.
 - **Increased appetite and weight gain** - steroids increase the appetite and often it is recommended that patients with MG visit a dietician.
 - **Susceptibility to infections** - the use of steroids slightly decreases resistance to infections.
 - **Osteoporosis** - steroids, when taken over a prolonged period, can make the bones more fragile by increasing calcium loss. It is recommended that patients take a diet rich in calcium and, especially where there is reduced sun exposure, supplementing calcium and Vitamin D may be necessary.
 - **Hyperglycemia or diabetes** - steroids make the body less capable of dealing with glucose and other sugars which may induce or exacerbate diabetes.
 - **Stomach upsets** - including indigestion, stomach burning or ulcers. Take steroids with food.
 - **Fluid retention** - this is caused by steroid effects on sodium and potassium metabolism. A salt-restricted, potassium-rich diet may help.
 - **Hypertension** - steroids may cause a rise in blood pressure.
 - **Skin changes** - the skin may bruise more easily or wounds may take longer to heal.

- **Changes in physical appearance** - changes may include swelling of the face, back of the neck and/or ankles. Acne, thinning of the skin and/or skin stretch lines may also occur.
- **Cataracts and worsening of glaucoma** - after prolonged use of steroids, cataracts or glaucoma may develop.
- **Alterations in hair growth** - darkening and/or increase in hair growth may occur but this disappears when the dose is decreased.
- **Stopping the medication too quickly** - can cause nausea, vomiting, pain, fever. Patients should never stop or change their dosage without their doctor's consent.

What Drugs Should Be Used with Caution?

Always consult with the doctor or pharmacist before taking any medication including prescription medicines, over-the-counter medications, herbs or vitamins.

The list provided below is not exhaustive and is not intended to replace professional advice. Each case is different and only the treating professional can advise in individual situations. Sometimes, it may be necessary to take medications on this list but this should only occur with close and informed medical surveillance. Always tell any other treating doctor that you have myasthenia.

Drugs that impair neuromuscular transmission and may increase weakness include (the names of specific drugs have not been included as these may change over time):

- **Antibiotics:** In general, the antibiotics most likely to cause problems are only available as intramuscular and intravenous preparations and therefore likely to be used only in a hospital setting or are only available as topical reparations (eg eye/ear drops, creams) and unlikely to cause problems as so little is absorbed into the body.
- **Cardiovascular Drugs:**
 - **Anti-arrhythmic** - should be avoided if possible.
 - **Beta Blockers** – there are many in this class, often identifiable by the 'olol' ending.
 - **Calcium Channel Blockers;**
 - **Some Cholesterol Lowering Drugs;**

- **Anti-Rheumatics** – should be avoided if possible. Quinine tablets and the quinine in tonic water also produce weakness in some myasthenics.
- **Anticonvulsants** – the overall risk of using anti-epileptics in MG is thought to be small.
- **Psychiatric Medications** – should be used with caution.
- **Anti-Spasmodic Drugs** – should be used with caution.
- **Ophthalmic Medications** – even though these drugs are used in drop form, they are absorbed into the circulation and can occasionally cause an increase in weakness.
- **General Anaesthetics:**
 - **Neuromuscular Blocking Drugs** - (known as muscle relaxants) can have a direct effect on the neuromuscular junction and, by blocking transmission, cause paralysis, including the muscles of respiration.
 - **Inhalation Anaesthetics** – may have a direct effect on the neuromuscular junction.
- **Botulinum Toxin** – directly affects the neuromuscular junction.
- **Narcotics** – should be used with caution. The use of Mestinon® can increase the effect of narcotics.
- **Magnesium** – the normal use of magnesium (eg laxatives, antacids, magnesium supplements) is unlikely to cause a problem. However, in renal failure, the magnesium levels can raise and cause muscle weakness. Magnesium sulphate is used intravenously in high doses for severe toxemia in pregnancy and can cause serious weakness.
- **X-ray Examinations with Contrast** – these agents are used to gain a better image in CT scans etc. The newer contrast agents are safe to use.
- **Vaccinations** – vaccines come in 'live' or 'inactivated' forms. Caution should be taken with the 'live' (the virus is live, but too weak to cause disease in people with normal immune responses) vaccines particularly if it is a first dose (ie. the patient will not have any pre-existing immunity). Inactivated vaccines are safe to use in myasthenia gravis, and may be very important in preventing complications

PART THREE:

LIVING WITH MYASTHENIA GRAVIS

What Important Factors Need To Be Considered in the Medical Management of Myasthenia Gravis?

In general, it is very important to keep myasthenia gravis in its place, and try not to let it take over one's life. However, there are certain precautions patients should take to ensure that they receive the best possible care. Any medical professionals, close friends and family should know the patient is taking steroids, immunosuppressants, having plasma exchange, IVIg etc. An identification card which provides details of necessary medications, doctor's name and phone number should be carried by all patients. Upon application, the MG Association of Queensland provides a Medi-Alert Card to its myasthenic members. This is valuable information should an emergency occur.

Other important issues to note are:

- **Medication regime** – it is important to take medication regularly and on time. Important information about taking:
 - **Mestinon® (pyridostigmine)** – it is important to take Mestinon on time and exactly as it has been prescribed. If one dose is missed within an hour of the prescribed time, the patient should take the missed dose and continue with other doses as scheduled. If the dose is missed by more than one hour, the patient should immediately take the dose and then wait the required 3 to 4 hours before taking the next dose. Subsequent doses should be taken with the prescribed intervals as well. Mestinon Timespan® is a higher dose and longer acting preparation that is often taken last thing at night.
 - **Steroids (prednisone/prednisolone)** – if a patient forgets to take steroids at the usual time but remembers later on the same day, the missed dose should be taken immediately. If the patient forgot to take yesterday's dose, just take the usual dose for the current day. If steroids are taken on an alternate day schedule and yesterday's dose was forgotten, then yesterday's dose should be taken today. Tomorrow resume the alternate day schedule.

- **Surgery and Anaesthetics** – patients should always make sure the anaesthetist is aware they have myasthenia gravis. MG used to pose challenges for anaesthetists. Nowadays, they are so well aware of them that it rarely causes problems. Local Anaesthetics are injected into the nerves to ‘freeze’ them by blocking their electrical conduction for several hours. This works the same whether the solution is injected near the nerve endings (eg into the jaw nerve in dentistry) or around the spinal cord roots (eg spinal and epidural anaesthetics). Spinal and epidural anaesthetics are suitable for many operations below the waist and are also better for people with MG should the type of surgery warrant it as it allows the patient’s MG treatment to continue normally and avoid the possibility of inadequate breathing caused by some general anaesthetics.

Before any *General Anaesthetic* it is vastly better if the myasthenia gravis is under the best possible control. This may mean tuning up with plasma exchange or IVIg a week or so before surgery. Surgery should then be as safe as it is for anyone else as long as anaesthetic care is taken over the following:

- **Muscle Relaxants** – these drugs paralyse all the voluntary muscles by blocking muscle ignition. They are given during surgery to help the surgeon to get easier access for ‘deep operations’. Because they also paralyse the breathing muscles the patient must be connected to a breathing machine (ventilator). Remember, however, that some operations do not need any muscle relaxants and the patient can breathe without any assistance.

With their lower reserve of muscle-triggering power, myasthenics are extra sensitive to muscle relaxants and need five or even ten times lower doses. The ensuing paralysis is usually stopped after the operation by injecting the short-acting neostigmine, which is a ‘cousin’ of Mestinon®.

- **Mestinon®** – patients should not stop taking Mestinon® before surgery but, as Mestinon® counteracts muscle relaxants, a slightly higher dose may be required.
- **Steroids** – the body naturally produces extra steroids as a result of stress. Long term steroid treatment reduces that response so it is usual to have extra steroids by injection before, during and after surgery in order to boost the body’s own efforts.

- **Dentistry** – prevention is vital to avoid dental emergencies as these can aggravate myasthenia gravis. Gums are liable for infections and, with immune-suppressants, infections are more likely and healing may take longer than expected. Also, weakness of the jaw muscles can affect the closing of your teeth and that, in turn, can create extra stress or even pain.

If the patient's myasthenia is under control, there is no reason why normal dental care cannot occur. The dentist needs to know what the patient's limitations are, and be prepared for them. Also, it is vital that the dentist consults with relevant specialists at the planning stage if surgery is necessary. If the patient has to have an anaesthetic, local anaesthetics are preferable to general ones. General anaesthetic should never be used outside the hospital setting.

- **Women's Issues** – many women notice their weakness is worse during the time of their monthly periods and others for a few months during menopause. There is no objection to Hormone Replacement Therapy (HRT) in patients with myasthenia gravis, nor to the use of the contraceptive pill.

Myasthenia Gravis very rarely affects the outcome of pregnancy as there is almost no extra risk of miscarriage or stillbirth. While MG occasionally gets worse during pregnancy, it more often does so for a few months afterwards. Prednisone and azathioprine should only be used in pregnancy when essential, although historical evidence suggests that they are not harmful to the unborn baby. Some other medications should definitely not be used during pregnancy and medical professionals will prescribe accordingly. With approximately one MG mother in eight, the newborn baby has a short-term weakness but they usually recover fully in the first three weeks or so. This 'neonatal myasthenia' is due to the transfer of the mother's damaging antibodies across the placenta. Unfortunately, the antibodies may also be transferred through the mother's milk so, if the baby is affected, breast-feeding should be avoided. Babies usually do not make anti-AChR antibodies of their own so they quickly recover as those transferred passively from the mother gradually decline. Very rarely, babies can have joint deformities (arthrogryposis). If so, this is because the mother's antibodies particularly attack (in utero) the baby's AChR which is slightly different from the adult's.

- **Allied Health Services:** Physiotherapists, dieticians, speech language pathologists, massage therapists can all provide useful information and treatment to help in the management of myasthenia gravis. As each person's needs are different, it is important to source these professionals and find out what they have to offer should you feel it may be worthwhile.

How Can Myasthenics Better Manage Living with This Rare Disorder?

Nobody wants to have a chronic long-term condition. However, a healthy way to live with one is to work at overcoming the physical and emotional problems caused by myasthenia gravis and to achieve the best possible physical capability and enjoyment out of life. Positive self-management on a daily basis is the key to living a healthy life.

Some handy hints to manage myasthenia gravis better include:

Managing Your Emotions: An early lesson that all myasthenics should learn is that myasthenia gravis and emotion are very poor bed fellows. Patients must accept they have the condition and learn to live with it and, at the same time, try to organise their lives so that they avoid situations leading to distress, anxiety or emotional crises. While recognising that not everyone will experience these, here are some commonly experienced emotions:

- **Anger** is one of the most common responses to chronic illness. People who have a chronic illness are often angry for having the illness, angry with family members and friends who might be unavailable when the patient needs them or who expect more of you than you can do or give. Recognising (or admitting) that you are angry and identifying why, or with whom, are important steps to learning how to manage your anger effectively.

There are several ways of helping to manage anger. Using “I” instead of “we” when expressing your feelings to others, modifying your expectations of yourself and others, and channelling your energy into new activities can help you manage your anger.

- **Depression** is a scary word and some people shy away from it by using terms such as “sad”, “feeling a bit down”, “finding it hard to cope at the moment”. Whatever it is called, depression is a normal reaction to chronic illness. While there are many signs of depression, there are several emotions that can lead to depression. These include a lack of joy, magnified sadness, fear or anxiety about the future, persistent pessimism, feelings of emptiness or isolation, frustration at not being able to do what you want or thought you could do, sleep disturbances, a feeling of loss of control over your life. Because the body and mind are so closely connected, depression can make your MG worse. Working to overcome depression is working to improve overall health. Depression feeds on depression so focus on your friends, your family, your goals, your pride in things you can do and

the power of positive thinking. Depression is not permanent and you and those who care for you and support you can help with its disappearance.

- **Stress** is a common problem for everyone, sick or well. If your body is not prepared to deal with the many demands put on it, stress occurs whether it be physical, mental, emotional or environmental. Everyone needs stress as part of their lives and, as long as it does not go past 'breaking point', stress can be helpful. Some warning signs of stress include biting your nails, pulling your hair, tapping your foot; grinding teeth and clenching the jaw; tension in the neck, head or shoulders; feelings of anxiousness, nervousness, irritability; frequent accidents or forgetting things you usually don't forget. If you catch yourself with any of the warning signs, take a deep breath and try to relax and think about what is making you tense. In order to deal with stress you must be able to identify the stressors and modify the situation. Problem solving is an important part of managing stress. Some simple ways to help manage stress include meditation, recreational reading, listening to music, visualisation/guided imagery, prayer, exercise, reading for inspiration or spiritual growth and hobbies.
- **Fear and Guilt** can have a negative effect on your emotions. If you feel guilty for having a chronic illness and live in dread of the future, you may be suffering more than need be. You can lighten your emotional burden by sharing your feelings in a support group or with others individually. Today, the internet is an amazing source of information and most organisations have a dedicated website.
- **Sleep Problems** can lead to fatigue or lack of concentration. Sleep is the time during which the body concentrates on healing. Only minimal amounts of energy are needed to maintain body functioning when we sleep. To help get a better night's sleep avoid eating and drinking alcohol just prior to going to bed, avoid caffeine late in the day, avoid eating foods with monosodium glutamate (MSG) late in the day, and avoid diet pills and diuretics unless essential. Develop a sleeping routine by setting up a regular rest and sleep schedule, exercise at regular times, get out in the sun for a short period every afternoon, (unless you are taking azathioprine), do the same things every night before going to bed (eg. reading a chapter of a book, having a warm bath) and use your bedroom only for sleeping.

If you fall asleep easily and are tired when you wake up in the morning, even after a full night's sleep, if you are overweight and if you snore a lot, then you may have a sleep disorder called Obstructive Sleep Apnea. This is a serious medical problem and can be life threatening. Anyone who finds they need more sleep now than before, or who regularly snore, should be evaluated by a medical specialist for sleep apnea or other sleep disorders.

Medications:

- No medication of any type should be taken without your doctor's knowledge. This includes MG medication. Always take medication with caution eg. never purchase cough mixtures off the shelf.
- Keep an ample supply of medication on hand to avoid the danger of running out, and ask your chemist to have some supplies in reserve for the times when MG medication is in short supply.
- For ease of mind, keep a small pill container with extra tablets in your purse, office desk, glove compartment of the car.
- Tape your medication schedule to the container you carry with you so that, if you have a problem in public, anyone trying to help you will have some idea of what your needs are if you are unable to speak clearly.
- If you need water during the night to take your medication but don't like to drink it warm, fill an insulated tumbler with ice for your bedside table. It will melt during the night and still be cool when needed.
- Some patients find having a small amount of food in their stomach (eg crackers) helps minimise some side effects when taking medication.
- If you take more than one medication or your doses change throughout the day, try counting out your day's medication into small plastic cups which can be labelled by number or hour. Your next dose will always be handy and ready at the proper time and will eliminate any doubt about your having taken the medication or not.
- Have written instructions about your medication in your home and at work and familiarise the people around you about where they are located. In case of emergency, they will know where to look.

Daily Living:

- Talking can be a problem for some myasthenics part or all of the time. Instead of using the telephone during these times, try using email or SMS.
- The same group of muscles involved in talking are also involved in eating. If you have difficulty chewing or swallowing, eat slowly, minimise your part in conversations and avoid hard-to-chew foods. Some myasthenics find it helpful to turn their head to one side when swallowing. This seems to change the position of the muscles to make swallowing easier. Adding a gravy, sauce, yoghurt etc to a meal often helps with swallowing. Some myasthenics also find it better to have six small meals rather than three big meals per day.
- As it is usual for myasthenics to tire as the day goes on, schedule important jobs to be done at home or at work for the beginning of the day when energy levels are usually higher.
- Listening and looking can become a problem if neck muscles fatigue easily. Try to position yourself in a strategic position to avoid unnecessary turning of the head and eyes.
- A boomerang pillow is great if you are confined to bed for any length of time. A neck rest pillow is also good when watching television or sitting for an extended period of time.
- An electric toothbrush is a great help when your arms or hands are weak.
- Smoking is harmful for myasthenics, especially if you have trouble breathing.
- Walk slower and you will probably walk further.
- Products in spray cans are usually poorly tolerated by myasthenics. The tiny particles conveyed by the propellant are absorbed into the bloodstream at an extremely fast rate. Also, beware of petrol fumes when filling the car.
- Take a look around the house to see if you are using unnecessary energy putting things that are used daily into cupboards. Save energy wherever you can.
- Learn to shop by phone. It can be done at your pace, save time and energy.
- Having a hot shower or bath can sap your energy. Keep the water temperature warm rather than hot.
- Soap-on-a-rope may be useful as it enables a person to place the soap around the neck while bathing. A hand-held shower rose, shower rails and a non-slip mat may also be of benefit.
- Heat and humidity brings another serious dimension to our MG with which we should learn to cope. Each person is different but probably the most common change is that, as the

body temperature rises, temporary worsening of MG occurs. Be aware of what symptoms worsen when it is hot, keep out of the sun, avoid exertion on hot or humid days, and try to work and exercise during the cooler part of the day. Also, drink plenty of water and avoid hot drinks.

- If walking a long way to the car when shopping is a problem, talk with your doctor about possible eligibility for a Disabled Sticker for car parking.

Travel:

- Talk to your doctor about your medical safety to drive a motor vehicle. There is a requirement in Queensland that any person who suffers from a listed chronic medical condition needs to notify the Transport Department accordingly and they need to produce a medical certificate indicating safety to drive.
- Airlines will provide wheelchairs to and from the planes if you request it at the time of booking your flights. You will be spared the long walk through the airport and you will enjoy your trip more.
- When travelling wear a medical identification emblem.
- In your travel wallet keep a list of current medications, medical history, allergies, a letter from your doctor listing your medications, doctor contact information and family contact information.
- Keep your medications with you, not packed in baggage that you check in. It is also a good idea to carry some medication on your person in case of emergency.
- A neck rest pillow is good for travelling as it supports a weak neck.

Parenthood and Myasthenia Gravis: If you are now, or plan to become, a new parent and you have MG, learn to take one day at a time and plan each day. Remember, the more strength you save, the more you can give your baby. Some hints to consider:

- Ensure that the baby's change table is at a height that allows you to stand straight and not have to bend at the waist.
- To bathe your baby, use a baby tub that slants and has a non-skid bottom. It helps keep baby's head out of the water and enables you to have both arms free for the bath.
- A portable crib can be used to transport baby from room to room. Keep it at arm's length if you feel weak.
- Think about a playpen. The baby can play, have room to move and even sleep in it and you can feel confident that the baby is safe.

- Keep baby's clothes simple, loose and easy to remove.
- Teach baby to hold you around the neck. This enables you to carry baby longer if your arms are weak.

Support Agencies: There is a wide range of providers and programs available to support patients with a chronic illness. Commonwealth Care Link Centres is a good starting point. Searching the internet can also be useful for meeting specific requirements (eg support programs run by different agencies).

Children with Myasthenia Gravis: Parents of children with MG should always think ahead in order to make their child's life as normal as possible and allow the child to be independent. Some suggestions include:

- Keep a food processor or stick blender handy if swallowing becomes a problem.
- When taking an outing go at the beginning of the day when the child is least tired.
- Give MG literature to all teachers at school, your child's friends' parents, relatives etc. and acquaint them with your child's needs and medication schedule.
- Get a letter from the doctor if your child needs to be excused from sport or physical activity at school.
- Purchase clothes that are easy to put on and take off.
- Keep dosages of medication separately so it is easy for your child to know what to take and when to take it. The child will start to recognise what medication to take and, when older, may be able to take the correct one upon awakening in the morning.

In Conclusion...

The current treatments for myasthenia gravis are sufficiently effective that the outlook for most patients is bright. Although there is no cure for MG, drug treatment has allowed individuals to show significant improvement in their muscle weakness and to lead relatively normal lives with a nearly normal life expectancy. In some cases, MG may go into remission, in which case the muscle weakness disappears. Remission may last as long as many years, and during these periods, treatment may not be necessary.

Ongoing research plays an important role in finding new answers and new treatments for myasthenia gravis.

GLOSSARY OF TERMS

What Does that Word Mean?

Acetylcholine (ACh) – is the chemical transmitter released from nerve endings on voluntary muscles. It is the 'ignition key'. It is far too small to be seen under any microscope.

Acetylcholine Receptor (AChR) – is the spot on the muscle which, when ACh binds to it, opens up channels into muscles to allow salt (Na^+) to enter and trigger the muscle into action. It is the 'ignition lock'. Like other large proteins, AChR's can just be seen under the most powerful microscopes.

Acetylcholine Esterase (AChE) – is a protein near the AChR's that destroys any spare ACh.

Anticholinesterases – are the drugs that block AChE so that any ACh lasts longer, giving it a better change of triggering. These drugs include Mestinon® (pyridostigmine) which is used for treatment and Tensilon (edrophonium) which is used for the diagnosis of MG.

Antibodies – are proteins specifically designed to destroy germs and block toxins. They are made by 'B cells' which come from the bone marrow and travel around in the blood and tissue fluids.

Autoimmune Diseases – are caused by cells or antibodies that can attack their own tissue or cell products.

Azathioprine (Imuran®) – is a drug that generally suppresses immune responses.

Benign – is the term used to describe a symptom that is of no danger to health; not recurrent or progressive; not malignant.

Bulbar – applies to the movements of chewing, swallowing, speech and breathing controlled by the lower brain stem.

Cholinergic Crisis – is usually brought on by prolonged too high a dose of Mestinon®, which can lead to respiratory failure.

Chronic – a long lasting condition as opposed to a short term (acute) condition. The term 'chronic' does not relate to the severity of the condition.

Congenital Myasthenia Gravis – strictly means MG that is there at birth but which may not be noticed until later in life. Many of the faults are in the AChR; others are in other genes at the neuromuscular junction.

Diplopia – double vision.

Diuretic – causing an increased output of urine.

Dysarthria – difficulty in getting words out. It is the physical movement of speech rather than finding the correct word in the brain (dysphasia) and is due to tongue or other mouth muscle weakness.

Dysphagia – difficulty in chewing and/or swallowing.

Dyspnoea – difficulty in breathing.

Electromyography (EMG) – where muscles are stimulated electrically, and the resulting electrical impulses are measured in the muscles they supply. Repetitive stimulation of nerve muscle may be used in the diagnosis of MG. It also helps the neurologist to differentiate between congenital MGs, and to differentiate LEMS from 'immune' MG.

Genes – molecular units of heredity that control and regulate biological functions

Hyperthyroidism – an excess of thyroid hormone resulting in an overactive thyroid gland.

Hypothyroidism – thyroid production is below normal resulting in abnormal thyroid balance.

Imuran® – see azathioprine

Intravenous Immunoglobulin (Intragam; IVIg) – slowly injecting into a vein the 'pooled' antibody fraction from normal blood. This procedure improves many autoimmune conditions.

Lambert-Eaton Myasthenia Gravis – is another form of autoimmune neuromuscular disease caused by antibodies acting against nerve endings. LEMS is rare and, while similar to the common form of myasthenia gravis, is different from it.

Mestinon® – is the commercial name for pyridostigmine. This drug is not a cure for myasthenia gravis but assists in managing the symptoms.

Muscles – are long tubes of protein woven together. When triggered, they shorten (contract), thus causing movement.

Mutation – an inherited or acquired change in the DNA sequence of a cell.

Neonatal Myasthenia Gravis – is the term used when myasthenia gravis in a newborn baby is caused by the passive transfer of antibodies from its mother.

Immune System - a complex system that is responsible for distinguishing us from everything foreign to us, and for protecting us against infections and foreign substances.

Nerves – these are two-way pathways. The afferent or sensory system relays electrical impulses from sense organs (eg eyes and skin) to the spinal cord and then the brain. The efferent or motor system relays from the spinal cord and cranial nerves to muscles and glands. The motor units relay the signals to muscles at special junctions (the neuro-muscular junction) and switch them either on or off. Sometimes, they act like dimmer switches, telling things to work harder or slower.

Ocular Myasthenia Gravis – is myasthenia gravis affecting only the eye movements and not other muscles. It does not affect individual eye focussing. That means that the eye can still 'see' but vision may be distorted or blurred or double because of paralysis or weakness of the eye movements.

Plasmapheresis or Plasma Exchange – is the method of cleaning the blood of unwanted antibodies to temporarily improve strength.

Prednisone, Prednisolone – are synthetic steroid drugs that generally suppress immune responses.

Ptosis – drooping or sagging eyelid/s

Synapse – any junction between a nerve and another nerve, a muscle or a gland. Signals can be passed either by chemical transmitters like ACh or by direct electrical triggering.

T Cells – are immune cells (from the thymus). Like antibodies, they also recognise foreign germs. They can either directly attack infected cells or recruit other cells to do that instead ('inflammation'). They are also needed to switch on 'B cells'.

Tensilon (edrophonium) – is a short-acting anti-AChE drug. It is used when diagnosing myasthenia gravis.

Thymus – the gland that produces immune T cells, especially before the age of 40 years, and exports them to the rest of the body. It is positioned between the breast bone and the heart and is important in autoimmune myasthenia gravis.

Thymectomy – is the removal of the thymus. This surgery seems to improve the myasthenia gravis in patients where there is a thymic tumour and also in patients where the onset has been at a young age (before 45 years).

Thymoma – a tumour on the thymus found in approximately 10% of myasthenics.

Vaccine – a germ or germ product made harmless. Still recognisable to T and B cells, it can be injected in advance, so stimulating these cells to multiply and forearm us before the real 'illness' comes along.

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