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 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 Myasthenia Gravis , 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 Myasthenia Gravis, a relative or a carer, you may find the science of Myasthenia Gravis 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 people with Myasthenia Gravis 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 **Myasthenia Gravis | healthdirect.gov.au)**

"Myasthenia Gravis is a rare disease of the neuromuscular system. Its main symptom is muscle weakness and it is caused by a breakdown in communication between the nerves and muscles."

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 can occur at any age.

When Was Myasthenia Gravis First Diagnosed?

MYASTHENIA GRAVIS 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, Myasthenia Gravis was not remorselessly progressive, but rather was fatigable – 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. Some modern Neuromuscular Blocking Agents had their base formulations derived from 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 Myasthenia Gravis 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 Myasthenia Gravis sufferers 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 Myasthenia Gravis 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 Myasthenia Gravis sufferers 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 Myasthenia Gravis sufferers. So, he proposed in 1960, that Myasthenia Gravis was an autoimmune disease also.

The above two paths finally came together in 1973. By then medical experts had realised that the ignition key, Acetylcholine (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 and Mycophenolate. It also provided a basis for treating Myasthenia Gravis with steroids. They were already in use for Myasthenia Gravis and, by the 1960's, it became clear that steroids 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 affected persons, when tested, prove AChR 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 different protein was causing neuromuscular inhibition. This protein is called Muscle Specific Tyrosine Kinase (MuSK) antibody.

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 receptors (AChR) on the fibres, thus making the muscle contract. Another chemical enzyme (acetylcholinesterase) then breaks down the acetylcholine, letting the muscle relax.

Myasthenia Gravis is an autoimmune disease. For unknown reasons, the body's own immune system produces antibodies against itself. If a person has Myasthenia Gravis, the antibodies the body produces damage the muscle receptor cells, reducing the number available on the muscle fibres. When this happens, the normal communication between the nerves and muscles is 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.

Presentation can be in different 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 Myasthenia Gravis diagnoses. 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 this may run in families. There are many subtypes and have different symptom presentations. Therefore, specialists use different forms of treatments in alleviating these symptoms.

- **Lambert-Eaton Myasthenic Syndrome (LEMS) –** Is a rarer cousin of Myasthenia Gravis. It is also an autoimmune disease but, in this case, the neuromuscular junction damage is done to the cells that produce acetylcholine. Thus, the message production is impaired. It also affects the voluntary muscles as well as some involuntary muscles which can produce other symptoms such as dry eyes and mouth, constipation. Unlike in Myasthenia Gravis, 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 mothers with Myasthenia Gravis 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 Myasthenia Gravis presentations directly affect sensation (eg sense of touch or temperature) or directly cause pain although aches in areas such as the back and neck can be caused by constantly trying to use weak muscles.

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;**
- **Weak, 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 muscles 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;**
- **Weakness in the shoulders and arms leading to difficulty lifting etc;**
- **Weakness in the legs leading to difficulty walking;**
- **General fatigue often brought on by physical activity.**

In Myasthenia Gravis muscle weakness becomes worse when the affected muscles are used and usually improves with rest. Symptoms may worsen as the day goes on but may get better after 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 menstrual period.

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

Who Gets Myasthenia Gravis?

MYASTHENIA GRAVIS presents at any age, in any gender or ethnicity. Under 40 years of age, females are affected with generalised Myasthenia Gravis twice as often as males. The incidence of Myasthenia Gravis in males increases over 50 years of age and males have a higher incidence of ocular Myasthenia Gravis. Prevalence appears to have risen over the past decades, probably from earlier diagnosis, better treatment pathways and increased lifespan of affected people.

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

Patients diagnosed with Myasthenia Gravis may have a potential to be predisposed to develop other autoimmune diseases e.g. diabetes, hyperthyroidism, rheumatoid arthritis, scleroderma and lupus. Some external risk factors to developing Myasthenia Gravis are suspected but yet unproven. Internal factors may include thymic tumours (‘thymomas’) which occur in approximately one patient in ten diagnosed 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 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 confirmed by health professionals such as Neurologists, Neuroimmunologists and Ophthalmologists who specialise in identifying and treating Myasthenia Gravis and its related conditions.

These practitioners will obtain a thorough patient account of symptoms and perform a general physical and neurological examination. Recognising the particular patterns of weakness gives the clues to the diagnosis. Once Myasthenia Gravis is suspected, it can be confirmed in several ways:

- **Observation of Muscle Weakness (Muscle Fatigue) –** The patient is asked to do repetitive movements of groups of muscles e.g. eyes, arms of legs and then tested for weakness. This is done without special equipment.
- **EMG (Electromyography) and SFEMG (Single Fibre Electromyography) –** Muscle 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 in 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 Myasthenia which may require different treatments.
- **Blood Test –** The most specific diagnostic test for Myasthenia Gravis. It is testing for typical antibodies which cause Myasthenia Gravis. Approximately 80% of all patients with Myasthenia Gravis have the Acetylcholine Receptor Antibody (AChR Ab). Another antibody tested for is the Anti-Muscle Specific Kinase (MuSK). If the blood tests confirm either of these antibodies, the doctor will term you Sero-positive; if they do not show up in the tests this is termed Sero-negative and this happens for approximately 10%-15% of all people diagnosed with Myasthenia Gravis. Recently, a further test may be

ordered by doctors – the Anti-Lipoprotein-Related Protein 4 (LPR4) for those who test negative for ACHR and MUSK antibodies.

- **CT Scan or MRI –** A scan or MRI of the chest is highly accurate in looking for thymic tissue. If it is present the doctors see whether it is overgrown or has developed a tumour. The tumours are usually benign but in rare cases a malignant growth may be found. These tumours are called thymomas. Chest radiography is relatively insensitive in screening for thymoma.
- **Tensilon Test –** Edrophonium®, a cousin of Mestinon®, is a short-acting anti-cholinesterase drug that is injected. Muscle 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 for Myasthenia Gravis .
- **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. This is not a formal test but is an indicator.

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 Myasthenia Gravis have a near-normal life expectancy.

Generally, Myasthenia Gravis is a persistent condition often requiring continuing treatment. Fluctuations in severity of symptoms can be the norm for some people. Muscle strength affected by Myasthenia Gravis for some time often recovers and the severity of the condition is usually modulated with the intensity of the treatment. Most patients can learn to live with Myasthenia Gravis and any treatment side effects to achieve a fulfilling life. Some even achieve remission (where no symptoms are experienced) with or without current treatment.

How is Myasthenia Gravis Treated?

DURING THE PAST SEVENTY YEARS, Myasthenia Gravis has gone from being an almost unknown fatal or disabling disease to current times in which the symptoms can be controlled so that now we have a Myasthenia Gravis community that includes many in the older age demographic.

There is a range of treatments available for treating Myasthenia Gravis, although often they are used in combination. The aims of treatment are firstly, to relieve the symptoms as much as possible and secondly, to treat the underlying cause with immunomodulating therapies that reduce the production of the autoimmune antibodies which cause Myasthenia Gravis. Thirdly, for some patients with identified abnormal thymic tissue, surgery is advised. 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.

- **Pyridostigmine (Mestinon®) –** This is usually the first-line treatment for Myasthenia Gravis. It is a reversible inhibitor of acetylcholinesterase so increases acetylcholine (ACh) stimulation of the remaining acetylcholine receptors (AChR). If there are insufficient acetylcholine receptors remaining to trigger the muscle action, the Mestinon® is unable to help. The use of Mestinon® does not cure Myasthenia Gravis or attack the rogue antibodies, but it assists the patient in coping by temporarily improving muscle strength with each dose.

  Mestinon® comes in 60mg and 10mg tablets, as well as a slow release 180mg dose which is usually used at night. Mestinon® assists the patient to temporally restore muscle strength in the muscles affected by Myasthenia Gravis. The dosage for one person may be very different from that required for another. Working with the relevant medical specialist requires a period of trial and response 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. Taking Mestinon® can sometimes cause stomach cramps and diarrhoea so taking it with food may help.

  Increasing the dose outside of recommendations in order to increase muscle strength may have the contrary effect and actually cause increased 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. If ongoing stomach cramps, diarrhoea, flickering eye movements or body cramps are experienced, the patient should consult their medical practitioner.

- **Immunosuppressants –** The principal drugs used to suppress the immune system in Myasthenia Gravis are a steroid such as prednisone and non steroidal immunosuppressants. Some common drugs used are **methotrexate**, **mycophenolate** and **azathioprine**. **Cyclophosphamide** and **cyclosporin** are sometimes used in severe Myasthenia Gravis if the symptoms do not respond well to conventional therapy. The response to these treatments can take weeks to many months, with the maximum effect taking months to years. By suppressing immunity generally, there may be the increased risk of infection. Medical specialists will often request patients have regular blood tests to monitor their health and any possible side effects.

- **Corticosteroids – Prednisone** and **prednisolone** are the most common steroids prescribed. Doctors may also use **hydrocortisone** and **dexamethasone** in certain circumstances. 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.

  Improvement in a patient's Myasthenia Gravis may occur in two to six weeks. Occasionally some initial deterioration in myasthenia symptoms can occur in the first few weeks of treatment so the dose is often increased slowly and with ongoing surveillance.

  Severity of Myasthenia Gravis, control of the symptoms and the development of adverse effects are all taken into consideration when the medical specialist determines the dosage of prednisolone. Trial and response are again used to determine the optimum maintenance dose, although during flare ups the medical specialist may change the dosage.

- **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 a surgical procedure. The number of plasmapheresis treatments needed depends on the protocol the physician has determined is best for each 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 'port' 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. You 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. Local anaesthetic may be offered. 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 you home.

Plasmapheresis works quickly to increase strength and most patients begin to improve after the first few days of the treatment. Reactions are rare and can be managed. 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 is done under close medical supervision.

- **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.

  Unlike plasmapheresis, it only requires a small temporary intravenous cannula and fluid line to be inserted, usually in the hand or arm. Sometimes, if finding veins to access is difficult or becomes difficult, the medical team may suggest a portacath be inserted.

  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. There are different brands of IVIg solution and your doctor will prescribe the best one for individual patients. The infusion clinic team will use an intravenous infusion pump to assist them in giving the IVIg accurately. It may take a few hours as the speed of administration depends on your 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. Patients sometimes get a headache, a rash or rise in blood pressure during the infusion so tell your infusion team if you do not feel well. If you become extremely unwell after your infusion do not hesitate to contact your medical team. Blood tests may be used to monitor individuals between infusions.

- **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 some people diagnosed with Myasthenia Gravis without significant thymoma. The thymus gland is composed of two lobes shaped roughly like a butterfly. It is located in the upper chest directly above the heart and between the lungs. Sometimes, thymic tissue may be found in other spots such as in the lower neck area.

  The surgery can be performed in several ways and the surgeon will determine the optimal procedure which may be based on position or size of the thymus. Convalescence after surgery depends on the procedure used and preoperative health. It can take up 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 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 subtituted 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 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.**

- **Immunosuppressants (Methotrexate, Mycophenolate, Azathioprine, Cyclophophasamide and Cyclosporin) –** These drugs suppress the immune system thus causing a reduction of the antibodies responsible for Myasthenia Gravis. Immunosuppressants are extremely useful as they allow smaller doses of other drugs, particularly steroids. Unfortunately, they may have side effects for some patients to differing degrees. Some side effects include gastrointestinal upsets, rashes and headaches. Issues with the liver and renal function and an increased risk of infections are monitored by regular blood tests and review by your medical practitioners. An increased risk of skin cancers can be mitigated by a commitment to being sun safe and having regular skin checks. If you have any concerns when taking these drugs please consult your medical team.

- **Rituximab –** This drug is used in the immunomodulating treatment pathway for Myasthenia Gravis. It is termed a Monoclonal Inhibitor CD20. This drug temporarilly removes and inhibits the regrowth of B cells which produce the antibodies which cause Myasthenia Gravis. It is useful for MUSK-MG and may occasionally be useful

for Myasthenia Gravis which has been very difficult to control on the previously mentioned medications. It is administered in hospital through an intravenous infusion. It may take a few weeks for you to feel a difference in your symptoms. For some individual requirements repeated infusions may be needed. Regular blood tests are recommended to monitor your health and any side effects from the drug.

The most common side effects occur at the time of the infusion, e.g. blood pressure and pulse fluctuations and allergy to the drug. You will be carefully monitored while the infusion is going. Rituximab may affect your immune response to infections. Consult you doctor promptly if you are feeling unwell or have an infection. Inform doctors that you have had Rituximab if you are requiring vaccinations or surgery.

- **Steroids –** Prednisolone and prednisone used in medicine are similar to those occurring naturally in the body.

  Adverse effects from the use of steroids is more concerning with very prolonged usage. They may include:

  - **Insomnia and mood changes –** 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 Myasthenia Gravis 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 with calcium and Vitamin D may be necessary. Consulting your doctor is recommended.
  - **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.

Your steroid medication should never be stopped or changed without your doctor's consent. If you are going to have a surgical operation or procedure ensure you inform your surgeon and anaesthetist.

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 supervision. Always tell any other treating doctor that you have Myasthenia Gravis.

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. Check with your doctor if the antibiotic being prescribed is contraindicated with your Myasthenia Gravis.
- **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.**
- **Anticonvulsants –** The overall risk of using anti-epileptics in Myasthenia Gravis 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 –** Affects Myasthenia Gravis symptoms.
- **Neuromuscular Blocking Drugs –** (Known as muscle relaxants) can have a direct effect on the neuromuscular junction.
- **Botulinum Toxin –** Directly affects the neuromuscular junction.

- **Narcotics and Strong Pain Medication –** Should be used with caution.
- **Magnesium –** The normal use of magnesium (e.g. laxatives, antacids,) is unlikely to cause a problem but care should still be taken. Supplements should be taken with your doctor's knowledge as they may cause increased weakness. Please be aware if required in a medical emergency Magnesium Sulphate may be used intravenously.
- **X-ray Examinations with Contrast –** These agents are used to gain a better image in CT scans etc. The newer contrast agents are generally 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.

The MGAQ has a resource on their website or by request which details a list of *"Drugs to be Used with Caution"* as an aid to assist you in conversation with your doctor in relation to prescribing drugs. Please visit **www.mgaq.org.au/drugs-and-mg**

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 if you are 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.

A list of medications that may worsen the symptoms of Myasthenia Gravis is available on the MGAQ website. Please visit **www.mgaq.org.au/drugs-and-mg**

A Drug Alert Card suitable to be carried in your wallet is available on application for financial members. The MGAQ strongly recommends the use of a medical alert wearable item. First responders are trained to look for such items. 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 you have Myasthenia Gravis. Myasthenia Gravis used to pose challenges for anaesthetists. Nowadays, they are so well aware of Myasthenia Gravis and how to care for patients with it that it rarely causes problems. Whether you are having a General, Epidural or Spinal or Local with/without sedation anaesthetic it is essential that you have a discussion with your anaesthetist before the procedure.

  During surgery which requires a General Anaesthetic, muscle relaxants are sometimes used. These muscle relaxants are drugs which paralyse (completely relax) the muscles to help the surgeon. In this instance they also paralyse the breathing muscles so the patient must be connected to a breathing machine (ventilator). People with Myasthenia Gravis may be extra sensitive and your surgical team will plan to take extra care of you. This may include a planned stay in an intensive care or high dependency unit after your surgery. The nursing team will be able to keep a closer eye on you than in the ward.

  It is not unusual for you to be scheduled IVIg or plasma exchange before your surgery to ensure the best possible control of your Myasthenia Gravis. Your anaesthetist will advise you about your medications to be taken on the day of surgery.

  - **Mestinon® –** Patients should not stop taking Mestinon® before surgery.
  - **Steroids –** The body naturally produces extra steroids as a result of stress. Long term steroid treatment reduces that response so it is common 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 Gravis 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 you have to have an anaesthetic, local anaesthetics are preferable to general ones. General anaesthetic should never be used outside a hospital setting.

- **Women's Issues –** Many women notice their weakness is worse during the time of their monthly period 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 Myasthenia Gravis occasionally gets worse during pregnancy, it more often does so for a few months afterwards. Some medications used in the treatment of Myasthenia Gravis should definitely not be used through pregnancy and breastfeeding. Your medical (Neurological, Obstetric and doctor) team will collaborate with modifying and prescribing your drug regime with your baby's welfare in mind.

  With approximately one Myasthenic mother in eight, the newborn baby has a short-term weakness but they usually recover fully in the first three weeks or so. Babies usually do not make anti-AChR antibodies of their own so they quickly recover as those transferred passively from the mother gradually decline. Breastfeeding is not usually contraindicated in Myasthenia Gravis. The mother's overall health, muscle fatigue, and drug treatment plan may impact breastfeeding.

- **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 People with Myasthenia 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 the 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 for those diagnosed with Myasthenia Gravis is that chronic or life changing disease and emotional health are poor bed fellows. Acceptance of life with the condition is a learned proficiency. Managing situations which may add anxiety or emotional stress may help. 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 you need 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 channeling 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 may make your Myasthenia Gravis worse. Working to overcome depression is working to improve overall health. Depression is not permanent. Self-help strategies and those who care for you can help with its resolution. Please consult your doctor who has a range of tools which can help.
- **Stress** is a common problem for everyone, sick or well. Everyone needs stress as part of their lives and, as long as it does not go past 'breaking point', stress can be helpful. Stress can occur from physical, mental, emotional or environmental causes. There are many signs of stress such as tension in the jaw, neck and shoulders, irritability and forgetfulness just to name a few. If you feel you are suffering any signs of stress, identifying the stressors is the first step. Once done, you can problem solve or modify the situation, both of which are important in helping to manage stress. Some simple ways to help the feelings of stress include meditation, recreational and inspirational reading, listening to music, visualisation/guided imagery, prayer, exercise, hobbies and having knowledge about your Myasthenia Gravis.
- **Fear and Guilt** can have a negative effect on your emotions. If you feel guilty for having an 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. The MGAQ website is an amazing source of information and access to dedicated closed discussion groups. The association is an excellent avenue to connect with others with Myasthenia Gravis in their lives. Please visit **www.mgaq.org.au**

- **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 function when we sleep. To help get a better night's sleep avoid drinking alcohol just prior to going to bed or caffeine late in the day. If you know foods which upset your stomach these are best avoided late in the day. Medications such as steroids and diuretics (unless specifically prescribed at night) are best taken in the morning to avoid a disturbed sleep. Develop a healthy sleeping routine. This can be done by setting up a regular activity, rest and sleep schedule. Get out in the sun for a short period each day if you can but be sun smart when you do. Do the same things every night before going to bed (eg. reading a chapter of a book, having a warm bath) and try to avoid using back-lit electronic devices before bedtime.

  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 Myasthenia Gravis 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 myasthenia 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 at room temperature, 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, using a tablet dispenser may help. Your pharmacist may be able to help you with dispensing your medication in a pre-packed time marked card. 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 people suffering with Myasthenia Gravis 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 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 gravy, sauce, yoghurt etc to a meal often helps with swallowing. Also, having six small meals rather than three big meals per day may be worth considering. Ask your doctor about a referral to a Speech Pathologist for assistance.
- As it is usual for people with Myasthenia Gravis 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. Timing your Mestinon® to coincide with activities which tire you is of benefit.

- 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, especially if you have trouble breathing.
- Walk more slowly and you will probably walk further.
- 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 or online. It can be done at your pace, saving time and energy.
- Heat is a trigger to many with Myasthenia Gravis. Having a hot shower or bath can sap your energy. Keep the water temperature warm rather than hot. Saunas, hot tubs/spas and heated pools should be used with caution. Safety when bathing on high symptomatic days is paramount. A hand-held shower rose, shower rails, shower chair and a non-slip mat may be of benefit. Having shampoos etc at chest height limits the need to bend or lift arms above the head.
- Heat and humidity bring another serious dimension to our Myasthenia Gravis with which we should learn to cope. Each person is different but probably the most common change is that, as the body temperature rises, myasthenic symptoms can temporarily worsen. A simple management plan is to keep out of the sun and heat and modify your activities on hot or humid days. Try to work or exercise during the cooler part of the day or in air conditioning only.
- Speak to your energy provider and your doctor about the possibility of electricity subsidies that can make energy bills more affordable.
- 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.
- Be aware that certain people find aerosols and certain fumes e.g. petrol, can aggravate their condition.

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 a wheelchair to and from the plane 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 (and when they are to be taken), 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 Myasthenia Gravis, 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. A discussion with your doctor to access these 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 Myasthenia Gravis should always think ahead in order to make their child's life as normal as possible and allow their 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 Myasthenia Gravis 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 waking 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 Myasthenia Gravis, 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, Myasthenia Gravis may go into remission, in which case the muscle weakness disappears. Remission may last for an extended time and treatment may not be necessary or be minimal. 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 Myasthenia Gravis .

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 Myasthenia Gravis 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 work 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 Myasthenia Gravis . It also helps the neurologist to differentiate between congenital Myasthenia Gravis, and to differentiate LEMS from 'immune' Myasthenia Gravis.

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.

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.

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.

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.

NOTES

Supported by

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