GUILLAIN BARRÉ SYNDROME

WHAT IS G.B.S.

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GBS

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Introduction

This guide has been written by neurologists and other specialists who have a particular interest in Guillain-Barré syndrome (GBS). It has to be honest and is meant to be reassuring. The information contained in this guide is an accurate and up to date account of GBS. Situations may arise in which you receive apparently conflicting opinions and information from different doctors and health care workers about various aspects of GBS. Unfortunately, the guide cannot respond in words to the conflicts or concerns that this information may cause. Consequently, if you do not understand or are worried by the information offered here, you must ask your medical specialist to explain. Don't be scared to quote from this guide if you feel intimidated or neglected! Any good doctor should be willing to listen and to explain.

What is GBS?

GBS is an uncommon illness causing weakness and loss of sensation that usually recovers completely after a few weeks or months. It is named after two French physicians, Guillain (pronounced Ghee-lane) and Barré (pronounced Bar-ray), who described it in 1916 in two soldiers who were affected by a paralysis but later recovered. New Zealand has one of the highest rates of GBS in the world and it affects about one person in 30,000 each year, ie 100-120 persons altogether each year in New Zealand. It can occur at any age from infancy onwards but is slightly more common in the old; it is more common in men than in women; it is not hereditary; it is neither passed onto children nor is it infectious and it is not caught from or transmitted to anybody else. However, it does often develop a week or two after a throat or intestinal infection.

What are the symptoms?

The first symptoms are usually either tingling (pins and needles) or loss of feeling (numbness) beginning in the toes and fingers. Legs feel heavy and wooden, arms feel limp and hands cannot grip or turn things properly. These symptoms may remain mild and clear up within a week or two without need for hospital admission but most people need to be admitted to hospital. At the earliest stage, it may

Can you tell me more about Miller Fisher syndrome?

About 5% of GBS sufferers have Miller Fisher syndrome (MFS) which was described in 1956 by Dr Miller Fisher. He described patients with paralysis of the eye muscles, in coordination of the limbs and loss of tendon reflexes but no weakness in the arms or legs.

Strictly speaking, that and only that, is MFS. The connection with GBS comes because some GBS patients have paralysed eye muscles too. Consequently, MFS and GBS can overlap.

Recently, special antibodies have been found in patients with MFS and in patients with GBS with eye paralysis but not in other GBS patients. These antibodies may be the cause of the eye muscle paralysis.

For more information about MFS, see our guide titled *Variants – Chronic and Acute*

Can I get a second attack of acute GBS?

The bad news is 'yes' but the good news is that the odds are against it; a figure of 3% has been estimated. This should not be confused with the chronic condition CIDP but some authorities do in fact reclassify people who have a second acute attack as having CIDP even though the second attack may have occurred many years after the first.

Notes

usually gets better on its own, a very important part of treatment is general nursing and medical care with physiotherapy and, if necessary, intensive care. No drugs have been proven to make any difference to the speed of recovery at this point in time, although further trials are being conducted in this area.

Can you tell me more about CIDP?

CIDP is less common than acute GBS (about 1:10) and most people reading this booklet need not bother with this section.

Like GBS, CIDP is an autoimmune disease of the peripheral nerves. Symptoms experienced by patients with both conditions are very similar.

CIDP is only distinguished from GBS by virtue of its pattern of progression. GBS is always defined if the low spot is reached within four weeks (and sometimes up to six weeks) although it typically happens within a few days. If the initial progressive phase lasts longer, and usually it is much longer, then the illness is called CIDP. Some CIDP patients are initially diagnosed with GBS and only when the deterioration continues over an extended period, or when one or more relapses occur after a period of improvement, is the illness reclassified as CIDP.

Although CIDP is a chronic condition, several different treatments are thought to be helpful. They all act by suppressing the damaging autoimmune response. Examples are steroids, azathioprine, plasma exchange and intravenous immunoglobulin. Obviously, suppressing the immune response cannot be undertaken lightly because it runs the risk of suppressing normal immune responses to infections. The decision whether to try these treatments has to be tailored by the doctor to the individual needs of each patient. However it is reassuring to know that demyelinated nerves can be repaired, that treatment is available and that some patients get better without treatment.

If you wish to know more about CIDP, see our guide titled *CIDP*.

be difficult for the patient to persuade the doctor that there is anything physically wrong. Within a few days it is all too obvious that something has gone wrong: legs simply will not bear weight, arms become very weak and the doctor finds that the tendon reflexes have disappeared.

How is GBS diagnosed?

The diagnosis of GBS is made from the clinical history (the story you tell your doctor) and medical examination, supported by laboratory tests. This means that the doctor will try to work out whether the history and clinical examination fit into the pattern of GBS. The doctor will particularly want to know of any recent possible infections or vaccinations, toxin exposure (such as insecticides or solvents), alcohol intake, tick bites, family history of nerve disease or symptoms of any coincidental illnesses such as diabetes (thirst, frequent urination, weight loss). Your answers to these questions might support the diagnosis of GBS or lead to a different diagnosis.

Investigations will normally include blood tests, a lumbar puncture and electromyogram (EMG). The lumbar puncture involves lying on one side and having a needle inserted under local anaesthesia between the vertebrae into the sac of cerebrospinal fluid that surrounds the nerve roots at the base of the spine. The idea is worse than the procedure really is and it does not usually hurt although there may be headache the following day. In most GBS patients, the cerebrospinal fluid contains much more protein than usual while the cell content remains normal. If different changes are found, the doctor has to review the diagnosis with even more care.

The electromyogram, or EMG, is an electrical recording of muscle activity and is a very important part of making the diagnosis of GBS. It is not done in all hospitals and may therefore require the patient to be transferred to a specialist unit where the test is available. If a nerve is stimulated with a brief electrical pulse (felt like a sharp tap or jolt), muscle activity can be recorded and the speed at which the nerve conducts electricity (the nerve conduction) can be worked out. Often in GBS, nerve conduction is slowed or even blocked altogether. The test usually lasts about half an hour. Some patients find the electrical stimulation rather uncomfortable but it is entirely harmless.

What happens next?

The worst degree of weakness is usually reached within two weeks and always within four weeks. Some patients deteriorate very rapidly to a state of severe paralysis over the course of a few days but this is uncommon.

The patient then enters a plateau phase that usually lasts a few days or weeks during which the course of the disease seems stationary. Most people are so weak during this stage that they are confined to a hospital bed where rest is probably a good thing. However, it is very important to keep all the joints moving through a full range to stop them stiffening up. The physiotherapist is in charge of this physical therapy and will be pleased to advise relatives and friends on what they can do to help.

Is GBS painful?

Unfortunately, some patients get a lot of pain during GBS, particularly in the spine and in the limbs. Other patients report GBS as an entirely painless experience, even when severely paralysed. Pain may come from the inflammation of the nerves themselves, from the muscles that have temporarily lost their nerve supply, from stiff joints, or simply because the patient is lying in an uncomfortable posture and is too weak to move into a more comfortable position. To combat the pain, the doctors will prescribe painkillers and the nurses and physiotherapists will help with repositioning and physical therapy. It helps to know that some pain is common in GBS. This pain should disappear as the condition improves and the occurrence of pain does not mean that anything else is going wrong.

Do patients need intensive care?

This subject and other items concerning GBS patients in intensive care are more fully detailed in our guide *Intensive Care*. A brief summary follows.

Since a patient with GBS can deteriorate rapidly, it is essential to treat him or her as a medical emergency initially. Once the progression of the illness is established, the doctors will be in a better position to judge whether or not the GBS patient will need to be admitted into an intensive care unit (ICU, sometimes called an intensive therapy unit or ITU). The remainder of this section is directed only towards the patients

the fluid or plasma. About 250ml of blood is removed at a time, the plasma is discarded and the blood cells are returned to the patient with clean plasma. The procedure is repeated several times on each of about five days until sufficient plasma has been exchanged. The risks of the procedure are extremely small and modern sterilisation has for practical purposes eliminated the risk of transmitting unpleasant infections in the clean plasma.

In other more recent trials, an alternative to plasma exchange has been discovered that is equally effective in speeding up recovery. This increasingly popular treatment is the infusion into a vein via a drip of a human blood product called gamma globulin or intravenous immunoglobulin (IVIG). This is given as a daily dose over three to five days. Put simply, IVIG is a cocktail of ‘good antibodies’ which fights off the ‘bad antibodies’ which are attacking the nerves. The administration of IVIG is simpler than plasma exchange and may be the preferred treatment in hospitals that have neither the plasma exchange equipment nor the expertise.

The above two treatments are probably not worthwhile in mildly affected patients, ie those who can still walk across a room unaided. If GBS patients cannot walk, or need help to walk, they should receive one of these treatments immediately the diagnosis is made (within 24-48 hrs at most).

The longer the delay in starting treatment, the less likely it is to be effective. On average, these treatments halve the duration of the illness in any individual case. They do not necessarily lead to an instant cure and some patients continue to get worse even on treatment. In these cases, all we can say is that the GBS patient in question would be even worse still without treatment. Some experts feel it is not worth giving any treatment after the first couple of weeks, unless the GBS patient is still deteriorating. Occasional patients require two courses of treatment. Although they do seem to shorten the duration of the illness, particularly the time on a ventilator and the time to walk unaided, they are a help rather than a cure and improved treatments are being sought. If you are worried that the expense or difficulty in prescribing or administering these treatments may result in their not being given, then ask your doctor why they are not being given. Remember that the cost of intensive care is also extremely expensive so that using these procedures actually saves money. Since GBS

Is there more than one type of GBS?

Yes. Perhaps it is a good idea to understand that GBS is a clinical syndrome (defined as an aggregate of symptoms) rather than a specific individual illness. In the majority of GBS cases, when the nerves become inflamed and demyelinated, the syndrome is due to 'acute inflammatory demyelinating poly[radiculo]neuropathy' or AIDP. Fortunately for GBS sufferers in this AIDP category, the part of the nerve attacked is the insulating sheath around nerves fibres termed myelin, equivalent to the plastic coating around electrical cables. This myelin sheath can be replaced by the myelin-forming cells, named Schwann cells, after Dr Schwann who described them.

Usually the conducting core of the nerve, equivalent to the copper core within electrical cables and called the axon, is not damaged. In the AMAN (acute motor axonal neuropathy) and AMSAN (acute motor and sensory axonal neuropathy) forms of GBS, the axons are damaged too. Although they can regrow, recovery takes longer and may be incomplete. Patients with AMAN or AMSAN may therefore make poor recoveries. More information can be read in our guide *Variants – Chronic and Acute.*

In some cases the illness may run a longer course than usual and become a chronic illness. This chronic version of the aforementioned AIDP is called CIDP (where C = chronic etc) and is described later in this booklet.

A variety of the acute condition is Miller Fisher syndrome (MFS) which is also described later. There are several other very rare conditions that are categorised as clinical variants of GBS; often they do not exhibit the full range of symptoms of the 'classic' description.

Is there a cure or any treatment for GBS?

Treatments for GBS have been evaluated in very large international studies involving many hundreds of GBS patients co-ordinated by teams of medical experts in the field. These studies are called 'Clinical Trials'.

Several of these trials have shown that, on average, plasma exchange is helpful for severely affected patients in the first week or two of the illness. Plasma exchange involves being connected to a machine that can separate the blood cells from

who are transferred to an ICU.

About 25% of GBS patients have weakness of the breathing, swallowing and coughing muscles and have to be placed on a machine that will take over their breathing called a ventilator or respirator. This process is called artificial ventilation. In addition to taking over the breathing, patients undergoing artificial ventilation have a tube placed in their throats, called an endotracheal tube, which prevents fluids in the mouth and acid in the stomach from 'going down the wrong way' into the lungs. If stomach acids find their way into the lungs they can cause severe damage and your doctors and other staff will do everything possible to prevent this from happening.

Admission to an ICU is less worrying than it sounds. Although occasionally GBS patients may be admitted to ICU for observation only, it is normally the case that patients on ICUs are placed on an artificial ventilator to take over their breathing. Under a short general anaesthetic, the connection to the ventilator is made to a tube placed in the windpipe (trachea) via the nose or mouth. This tube, the endotracheal tube, can be left in place for a week or two. If artificial ventilation is required for longer, a surgeon may make a small opening, called a tracheostomy, into the windpipe at the base of the throat, just below the 'Adam's apple'. This is more comfortable for the patient and permits artificial ventilation for as long as necessary. The tracheostomy is also performed under a general anaesthetic. Fortunately, in GBS, artificial ventilation is rarely necessary for more than a few weeks and the majority of patients do not need artificial ventilation at all.

When ventilation is no longer needed, the tracheostomy tube can be removed quite painlessly. The wound closes in a few days and eventually leaves a small scar below the line of the collar.

Intensive care in recent years has become a very sophisticated part of medicine that has enormously improved the care of severe GBS. To make this possible, pulse, blood pressure, temperature and blood chemistry have to be measured often. The pulse will be recorded by monitoring the heart beat (electrocardiogram) on a video monitor to detect abnormalities that may need treatment. Patients may need infusions into veins to provide fluids and give drugs. A tube called a catheter is placed in the bladder to drain the urine.

Another tube, called a nasogastric tube, may be passed through the nose into the stomach to provide nutrition because swallowing will be impossible. Constipation can be a troublesome problem at first but eventually nurses and patients invariably work out a regime of laxatives and suppositories that works.

Communication can be a problem for a patient who is unable to talk but with winks, nods, communication cards (usually available from the Hospital or otherwise contact the Support Group National Co-ordinator) and, above all, patience it is usually possible to get the message across. If the intensive care regime seems tedious, it is worth remembering that modern intensive care has reduced the mortality rate of GBS considerably. Fortunately, death from GBS is now a rare event, occurring in around 3% of cases. Death tends to occur more commonly in elderly people severely affected by GBS and with other medical illnesses such as heart, lung or kidney disease. Like any other illness, unexpected complications can arise. Death is more likely to be a result of a complication rather than GBS itself.

How long does it take to recover?

Eventually the numbness begins to recede and strength begins to come back. Once it is clear that this is a genuine improvement rather than wishful thinking, there is some cause for cautious rejoicing because improvement is likely to continue steadily. About 80% of the patients recover completely in that they are up and about walking within one year, and often much earlier than this. The time taken for recovery to occur is very variable. Sometimes it is only a week or two but most people remain affected for between three and six months.

The patients who do not recover completely may be left with minor degrees of weakness, numbness and sometimes discomfort that do not seriously interfere with their lives. A few however are left so disabled that they cannot resume their former occupations. This is usually because of residual weakness of their arms and legs so that manual work and walking are impaired. It is uncommon to be left dependent on a wheelchair for life but this unfortunately does occur in some cases. Improvement is fastest during the first few months but some patients report continued gradual improvement even after a year or two has elapsed.

What causes GBS?

The disease is due to inflammation of the peripheral nerves, often termed ‘neuritis’. It is like an ‘-itis’ anywhere else in your body: an angry redness and swelling that stops the organ in question from working properly. For example, laryngitis (inflammation of the larynx) leads to the loss of voice. The peripheral nerves are like the electrical cables around your house. They connect the central nervous system (ie the ‘mains’) to the muscles and to the sense organs in the joints and skin (ie the ‘appliances’). When these cables are damaged or cut, the appliances stop working because they have no electrical power, although are in themselves undamaged. Because many nerves are inflamed, GBS is called a ‘polyneuritis’. The most likely explanation for the inflammation is that immune cells called lymphocytes start attacking the nerves in error, instead of concentrating their energies on fighting off infections. This mistake in the immune system is an own goal you could do without! It is believed that the immune system has been tricked into making this mistake by an infection that often precedes GBS. Eventually the immune system realises its mistake and corrects it by either killing off the renegade lymphocytes or discharging them from the front lines of its army, thus stopping the attack on the nerves. A disease in which the immune system attacks its host’s own body is called an autoimmune disease and GBS is one of many diseases affecting the nervous system in this category.

For more general information, see our guide *Peripheral Nerve Disorders.*