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?

Guillain-Barrè syndrome is an autoimmune condition in which the person's peripheral nerves are attacked by the body's own immune defence system. As a result of the attack, the nerve insulation (myelin) and sometimes even the inner covered part of the nerve (axon) is damaged and signals are delayed or otherwise changed, causing paralysis and muscular weakness of the limbs.

The syndrome appears to be triggered by acute viral or bacterial illnesses, such as respiratory or gastrointestinal infections, occurring one to three weeks earlier. However, other events such as pregnancy, dengue fever, surgical procedures, insect bites and Bell's palsy have also been shown to be a cause.
GBS is named after two French physicians, Guillain (pronounced Ghee-lan) and Barré (pronounced Bar-ray), who described it in 1916 in two soldiers who were affected by a paralysis but later recovered.

It can occur at any age from infancy onwards; it affects both men and 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. There is no cure for GBS, but treatment can reduce the severity of your symptoms and shorten the duration of the illness.

**WHAT ARE THE SYMPTOMS?**

It can present in many different ways making it difficult to diagnose in the early stages. First symptoms include varying degrees of weakness or tingling sensations in the legs, or in some instances may begin as trouble moving your eyes or face. In many cases this spreads to the upper body and in severe cases leads to almost total paralysis. GBS can be dangerous if the muscle weakness affects the chest muscles responsible for breathing.

The symptoms can occur quite rapidly over hours to days or slowly over weeks. Most patients reach peak weakness within the first 4 weeks.

**The symptoms of Guillain-Barré include:**

- muscle weakness in your legs that travels upward and worsens over time
- tendon reflexes have disappeared
- difficulty walking steadily
- difficulty moving your eyes or face, talking, chewing, or swallowing
- severe lower back pain
- fast heart rate
- difficulty breathing
- paralysis
THE PERIPHERAL NERVES

The peripheral nervous system is a network of 43 pairs of motor and sensory nerves that connect the brain and spinal cord (the central nervous system) to the entire human body. To the right is a drawing of the peripheral nervous system.

HOW IS GBS DIAGNOSED?

Guillain-Barré is difficult to diagnose at first. This is because the symptoms are very similar to those of other neurological disorders or conditions that affect the nervous system. Your doctor will ask questions about specific symptoms and your medical history.

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.
The following tests are used to help confirm a diagnosis:

**Spinal Tap/ Lumbar Puncture**

A spinal tap also referred to as a lumbar puncture is a medical procedure which 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. In most GBS patients, the cerebrospinal fluid (CSF) 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.

**Electromyography (EMG)**

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.
WHAT HAPPENS NEXT?

The worst degree of weakness is usually reached within four weeks and always within six weeks. Some patients deteriorate very rapidly to a state of severe paralysis over the course of a few days but if diagnosed quickly this might be avoided.

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 THERE A CURE OR ANY TREATMENT FOR GBS?

Everybody with Guillain-Barré should be admitted to a hospital for close observation. The symptoms can quickly worsen and can be fatal if they aren’t treated. In severe cases, people with Guillain-Barré can develop full-body paralysis. Guillain-Barré can be life-threatening if paralysis affects the diaphragm or chest muscles, preventing normal breathing.

Guillain-Barré can’t be cured. The goal of treatment is to lessen the severity of your symptoms and keep your body functioning while your nervous system recovers.

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’.
Plasmapheresis / Plasma Exchange

Plasma exchange is helpful for severely affected patients in the first week or two of the illness.

Plasmapheresis is intended to remove the antibodies attacking the nerves from your blood. Plasma exchange involves being connected to a machine that can separate the blood cells from 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 for 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.

Intravenous immunoglobulin (IVIg)

IVIg is only licensed for use in the treatment of GBS or (AIDP).

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 IVIg made from the ‘plasma’ of donated human blood.
‘Plasma’ is the clear fluid part of the blood. Currently IVIg is only given in hospital, usually in a day care unit. It is given through a drip and the rate, dose and time are calculated individually for each patient.

If the treatment is successful it may have to be given on repeated occasions 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.

**When to have these treatments:**

The above two treatments are probably not worthwhile in mildly affected patients, i.e 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 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.

**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). The remainder of this section is directed only towards the patients 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. This is more comfortable for the patient and permits artificial ventilation for as long as necessary.

The tracheostomy is 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

Communication can be a problem for a patient who is unable to talk but with winks, nods, communication cards and, above all, patience it is usually possible to get the message across. Contact us if you need any communication cards.

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 1 in 15 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.

REHABILITATION IS VITAL FOR RECOVERY.

The length of time you have GBS is unpredictable, and may require months of hospital care and rehabilitation.

As nerve function returns, patient may need assistance to learn how to use affected muscles. Rehabilitation may include several types of therapy:

**Physical Therapy:** This stimulates muscles and joints to rebuild strength, flexibility and range of motion.
**Occupational Therapy**: focuses on activities to help patient be as self-sufficient as possible in daily life.

**Assistive devices**: Patient may need to learn to use assistive devices, such as leg or arm braces, canes, walkers and wheelchairs to aid mobility during recovery or, if GBS causes permanent disabilities, for long-term use.

There are also multiple alternative therapies that have a variety of results and differs person to person. Therapies such as water, music, acupuncture and many more.

**Physiotherapy treatment for GBS will:**

- Regain patient’s independence with everyday tasks.
- Retrain normal movement patterns by teaching patient how to achieve activities in different ways.
- Muscle strength training, exercising as often as possible in the correct way.
- Stretch tight muscles and prevent soft tissue contractures.
- Improve patient’s posture in lying, sitting and standing and sleeping.
- Increase patient’s mobility.
- Increase balance and coordination.
- Increase fitness and energy levels.
- Increased ability to relax.
- Promote recovery.

**WHAT CAUSES GBS?**

The precise cause of Guillain-Barré is unknown.

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, the peripheral nerves are like the electrical cables around your house. They connect the central nervous system (i.e., the ‘mains’) to the muscles and to the sense organs in the joints and skin (i.e., 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.

In extremely rare cases, people can develop the disorder days or weeks after receiving a vaccination. There are systems in place to monitor the safety of vaccines, detect early warning signs of side effects, and record any cases of Guillain-Barré that develop following a vaccination.
Campylobacter jejuni infection has been associated with Guillain-Barré. Campylobacter is one of the most common bacterial cause of diarrhea in the United States. It’s also the most common risk factor for Guillain-Barré. Campylobacter is often found in undercooked food, especially poultry.
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.

Acute inflammatory demyelinating polyneuropathy (AIDP):

The most common form of GBS, and the term is often used synonymously with GBS. It is caused by an auto-immune response directed against Schwann cell membranes. In the majority of GBS cases, when the nerves become inflamed and demyelinated, the syndrome is due to 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, called the axon, is not damaged.
In some cases the illness may run a longer course than usual and become a chronic illness. This chronic version of the afore mentioned AIDP is called CIDP (where C = chronic). Some CIDP patients are initially diagnosed with GBS and only when the deterioration continues over an extended period, or when 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 IVIg.

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.
**Acute motor axonal neuropathy (AMAN):**

AMAN attacks motor nodes of Ranvier (see diagram). It is probably due to an auto-immune response directed against the axoplasm of peripheral nerves. The disease may be seasonal and recovery can be rapid. Anti-GD1a antibodies are present. Anti-GD3 antibodies are found more frequently in AMAN.

**Acute motor sensory axonal neuropathy (AMSAN)**

Similar to AMAN but also affects sensory nerves with severe axonal damage. Like AMAN, it is probably due to an auto-immune response directed against the axoplasm of peripheral nerves. Recovery is slow and often incomplete.

In the AMAN and AMSAN 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.

**Miller Fisher syndrome (MFS)**

A rare variant of GBS acute disease and manifests as a descending paralysis, proceeding in the reverse order of the more common form of GBS. It usually affects the eye muscles first and presents with the triad of ophthalmoplegia (paralysis of the muscles within or surrounding the eye) ataxia, and areflexia.
Anti-GQ1b antibodies are present in 90% of cases. About 5% of GBS sufferers have Miller Fisher syndrome (MFS) which was described in 1956 by Dr Miller Fisher.

**Acute panautonomic neuropathy**

The most rare variant of GBS, sometimes accompanied by encephalopathy (disorder or disease of the brain). It is associated with a high mortality rate, owing to cardiovascular involvement, and associated dysrhythmias (abnormality in a physiological rhythm usually in brain or the heart).

Impaired sweating, lack of tear formation, photophobia, dryness of nasal and oral mucosa, itching and peeling of skin, nausea, dysphagia, constipation unrelieved by laxatives or alternating with diarrhea occur frequently in this patient group.

**Bickerstaff’s brainstem encephalitis (BBE)**

A further variant of Guillain–Barré syndrome. It is characterized by acute onset of paralysis of the muscles within or surrounding the eye, ataxia, disturbance of consciousness, hyperreflexia (overactive or overresponsive reflexes) or Babinski’s sign.

The course of the disease can be a single phase or remitting-relapsing. Large, irregular hyperintense lesions located mainly in the brainstem.
BBE despite severe initial presentation usually has a good prognosis. Magnetic resonance imaging (MRI) plays a critical role in the diagnosis of BBE. A considerable number of BBE patients have associated axonal Guillain–Barré syndrome, indicative that the two disorders are closely related and form a continuous spectrum.

**Other Variants:**

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.

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**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.