



# **Guillain-Barré syndrome**

## **& the associated acute**

### **variants**

Helpline: 0800 374803 (UK) 1800 806152 (ROI)

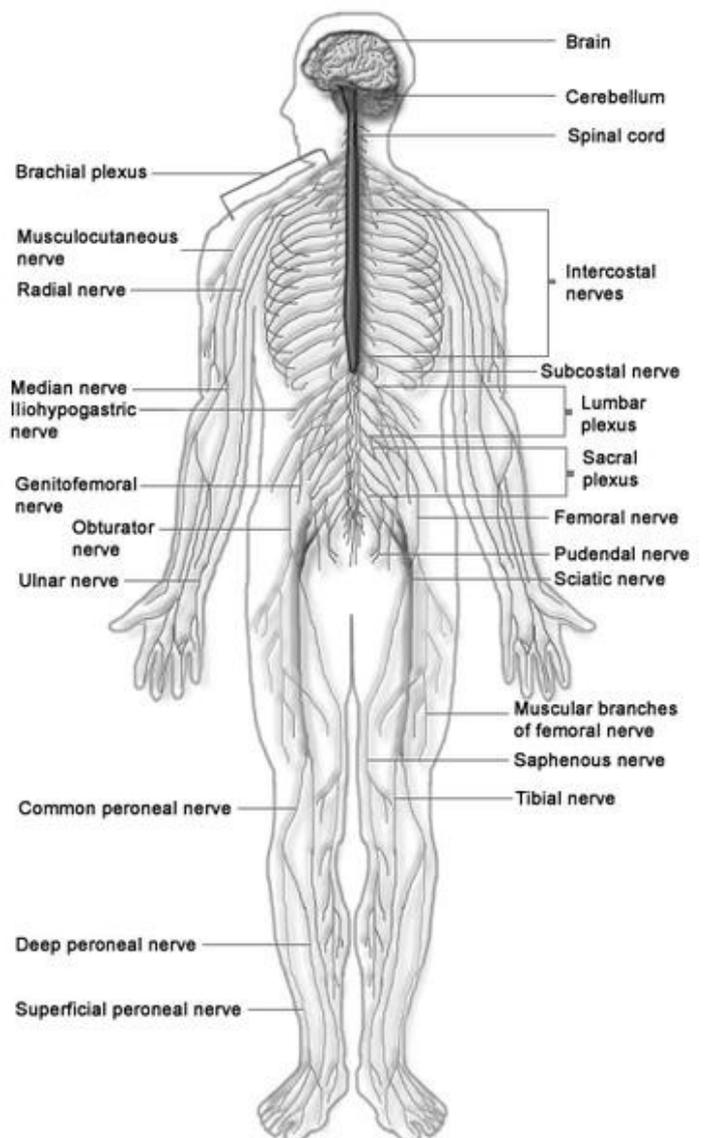


This guide has been written by neurologists and other specialists who have a particular interest in Guillain-Barré syndrome (GBS). It has to be honest and is meant to be reassuring. The information contained in this guide is an accurate and up to date account of GBS. Situations may arise in which you 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, or almost completely, after a few weeks or months. It is named after two French physicians, Guillain (pronounced Ghee- lan) and Barré (pronounced Bar-ray). In 1916 they described two soldiers who were affected by a paralysis but later recovered. It affects about one person in 40,000 each year, ie 1,500 persons altogether each year in the United Kingdom. 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 gut 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 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 spinal fluid (or CSF) 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 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.

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.

Occasionally the diagnosis can be delayed by a few hours or a couple of days whilst results of tests come back when there are atypical features. Sometimes doctors elect to treat before they are certain of the diagnosis to be on the safe side.

### **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 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?**

Around a quarter of GBS sufferers are admitted to intensive care units (ICUs, sometimes called intensive therapy units or ITUs) for special care if their illness is judged severe or moderately severe. Admission to ICU is particularly recommended for patients with weakness of their breathing, swallowing or coughing muscles. A machine called a ventilator will be

introduced to take over their breathing function and to stop fluid and secretions from slipping down the throat into the lungs where infection and lung damage may arise.

### **What is an intensive care unit?**

This is a special unit within hospitals, staffed by medical support personnel who are specially trained in the high levels of care required by each patient. There is nothing sinister or depressing about these units. On the contrary, they are busy and cheerful places where patients are under constant watch, day and night, and everything is done to ensure that the patient receives the highest level of care possible. At first sight, there appears to be a daunting amount of equipment at the bedside, but you will be surprised how soon you and your affected relative or friend can come to understand the function of each piece of machinery.

### **Why has the patient been admitted to the unit?**

The patient's breathing needs to be supported. This is done by attachment to a ventilator. A ventilator is a sophisticated machine which will simulate the patient's own breathing requirement while he/she is temporarily unable to breathe unaided. This may be done via the nose or mouth in the shorter term, or for those patients who are likely to require ventilation for more than a short period, then via a tube placed through a small incision in the neck (tracheostomy). This small opening is quite comfortable and will be closed up as soon as the patient is able to breathe again without assistance from a ventilator.

The patient's immediate reaction to being put on a ventilator is frequently one of relief that the struggle to breathe normally is now over. The patient's heart will be monitored on a screen to watch for any irregularities. A thin tube (catheter) may be used to drain urine from the bladder. In order to feed the patient whose swallowing ability is impaired by the GBS or made impossible by the plastic breathing tube, a special tube called a nasogastric tube will be passed through the nose and down the throat and oesophagus into the stomach so that liquid food may be taken in.

Pulse, blood pressure, temperature and other vital signs will be regularly monitored. Airways and lungs will be kept clear by a method of suctioning, as and when required. This is an essential procedure which, when completed, gives the patient considerable relief. However, it is noisy and if visitors find

this distressing or unpleasant they should quietly leave the unit until it is over.

This all sounds a bit frightening but remember that these procedures are all regularly used in ICUs and are essential for the patient's wellbeing. Each support mechanism will be discarded as the GBS sufferer improves and they are no longer needed.

### **How does the ventilated patient feel?**

At first, patients are very alarmed at the new situation and surroundings in which they find themselves. However, they soon become familiar with what is going on and begin to understand the routine. A simple but careful explanation is essential to put the patient's mind at rest.

The first thing to note is that the GBS patient cannot speak and may also have a reduced or absent sense of taste and smell. Some patients will also experience visual disturbance. Hearing is rarely impeded, so the patient can generally understand and acknowledge all that is going on.

However, appreciation of the surroundings may also be dampened by sedative or painkilling drugs which are often used to make GBS patients more comfortable.

Some patients do experience an increase in skin sensitivity so although touch is important, care must be taken. In some rare cases even a light touch may cause very severe pain which the patient cannot easily communicate to you.

GBS is a paralysing illness. Paralysis is temporary but can be quite extensive and the patient is fully aware of the lack of movement. This can be perplexing, frightening and hard to accept.

The GBS sufferer, although receiving the requisite amount of nourishment, may lose weight. This is the result of wasting of the muscles and cannot be avoided. Pain may be experienced to a greater or lesser degree at various sites around the body, for which appropriate medication will be given. The level of any pain must always be borne in mind when moving the patient and utmost care taken to ensure that all movements are carried out as gently as possible.

During the severe phase of the illness, GBS patients can go through hot and cold spells and will frequently request a fan to be turned on and off.

Hallucinations, unusually vivid daydreams or nightmares, are not uncommon for ventilated GBS patients. They may be worsened by sedative or painkilling drugs but can also arise in patients without any drug effects. They are not necessarily frightening but the patient may be convinced of their reality.

Many GBS patients are alert and acutely aware of what is going on. They feel vulnerable, isolated and locked-up inside their illness. Considerable frustration occurs because they are unable to talk whilst on the ventilator, and you may encounter some irrational or uncharacteristic behaviour.

It is never easy for them to come to terms with what has happened, so do not be surprised if they are variously tearful, bad tempered or panicky.

Everyone coming into contact with the ventilated GBS patient should remember at all times that the patient is quite aware of their reliance upon the machinery to which they are attached.

Remember too that from a mental and emotional standpoint, loss of movement and inability to speak makes patients feel fragile and vulnerable. A less than caring action or unsympathetic attitude can set the alarm bells ringing inside the silent patient.

### **What can family and friends do to help**

A great deal!

Your first task is to understand, at least in outline, what this illness means. Speak to the doctor in charge of the case as soon as you can to get yourself into the picture. Some doctors are better than others at explanations. Don't hesitate to ask questions. Have you read the guide published by **GAIN**? Does the hospital know that there is a national charity?

Secondly, familiarise yourself as soon as you can with the ICU. Get to know the regular nursing staff who will give you a daily update on progress. If a patient is to receive a new treatment or procedure, make sure he/she knows about this in advance and understands why it is being undertaken.

A physiotherapist may begin passive movement of the limbs whilst the patient is bed-bound. Get to know the physio and keep yourself updated on procedure and progress. There is a lot he/she can tell you.

The patient cannot talk but is anxious to communicate. Make sure the speech therapist is involved in advising on communication aids. If good

facial strength has been retained, then lip reading will be effective. Some patients retain finger movement and can write letters in the air or on the palm of the hand. A common method of communication with a patient whose movements are restricted to the eyelids, is to use a question and answer technique with the patient answering with one blink for 'yes' and two for 'no', sometimes running through the alphabet until the correct letter is found. This can be improved upon by pointing to the letters on an alphabet board and asking 'Is it on this line? Is this the letter?', responding as before. If the patient is strong enough, he/she may be able to point at an alphabet board with a finger or pointer attached with a headband. A very useful method of communication is by the use of communication cards which pre-empt many questions or comments the ventilated patient is likely to make. Copies are available from **GAIN**.

The GBS patient puts a lot of effort in trying to communicate and you soon find a method that works and you will become quite expert. Encourage others to understand too.

The GBS patient is socially isolated and needs to be stimulated. Make sure he/she knows the day of the week and the date. Encourage friends and family to send cards and write letters about what they are up to. If the patient cannot see TV, relate what is going on in the outside world. Read extracts from a national or local newspaper. Would the patient like an audio book played?

Always include the patient in bedside conversation.

Financial worries may be bothering the patient, especially if he/she is a breadwinner. Get in touch with the Social Worker at the hospital who will advise on State benefits and claims. Alternatively, your local Citizens' Advice Bureau dispenses free and expert advice on benefits. **GAIN** may also be able to help with travel costs for visiting family.

Early action is essential as many benefits cannot be claimed retrospectively. Inform the patient's employers about GBS and confirm the situation on job security. Patients worry a good deal about such matters.

It is advisable not to eat or drink (or refer to such things) in front of the patient as this can be deeply intimidating for someone who can do neither.

The patient's view of the world may be very dull, so it helps if you wear bright cheerful colours and ensure that flowers (if they are allowed) and 'get well'

cards can be seen.

GBS patients tire easily, so do not encourage lengthy visits. At the end of each day, as evening approaches, make sure you leave the patient in the best possible frame of mind, ready for the night. Have you turned off or turned down the radio? Has the patient all he/she needs for the night, such as the fitting of night splints (if required)? Can the patient easily attract the nurse's attention? Paralysed patients are especially afraid of the night; so ensure they are in a calm mental state before you depart.

Because patients have a small span of attention and may be on sedative drugs, they tend to drop off to sleep quite frequently, though perhaps only for a nap. Alternatively, they may appear to be half awake but inattentive. This could be because they are hallucinating. This is nothing to worry about and you should continue to talk to the patient and try to regain their attention.

Try to understand the patient's pain and the frequency and type of medication being given to alleviate it. Some GBS patients experience very little pain, but if it is a factor then it is a comfort to know that it will decrease alongside the patient's general improvement.

Remember too that the patient's morale is at a low point. The little things you can do will mean a lot. Does the patient need a hair wash? Do nails need manicuring? Can you help by massaging the patient's hands or feet? Is the patient having problems causing any anxiety over his/her relationship with the medical staff? If so, you must resolve such difficulties speedily.

As the patient's breathing improves, he or she will be gradually taken off the ventilator, starting with just a few minutes and building up from there.

Patients can get quite panicky at the beginning of this procedure as they have become reliant on the ventilator and do not believe, initially, that they can breathe again without it. Reassure them that their natural ability to breathe is returning and that this is the start of getting well.

### **In summary**

It is impossible to cover every single aspect of the GBS patient in the ICU. This is a very personal illness and each patient has his or her particular set of problems and worries to cope with. Your role is to offer love, comfort and reassurance during this difficult period. To do this effectively, you must remain calm and resolute and give constant encouragement on progress. Patients easily lose sight of how they are doing so keep yourself well

informed by the medical staff. Writing a diary of daily events will help you keep a perspective on progress. You can relate this to the patient who may not realise how he/she is getting on.

Some days are better than others for GBS patients and it is hard to be a hero every day, but you must keep up a constant flow of encouragement.

For the close family, this period of the illness is quite stressful, so don't forget to look after yourself and stay well.

### **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.

One lasting problem is fatigue. Doctors are not good at appreciating, measuring or treating fatigue. Sometimes this lasts longer than weakness but pausing the return to normal life and not overdoing activity too fast usually helps fatigue in the longer term.

### **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*.

### **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 lose their insulation (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 later in this guide

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 and in the booklet *CIDP and associated chronic variants*.

A variety of the acute condition is Miller Fisher syndrome (MFS) which is 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.

### **Acute Variants**

**Axonal GBS** - It had been known for some time that in severe cases of GBS, a 'bystander' effect of the demyelination of the nerve could be damage to the nerve core or axon. In 1986, Feasby et al<sup>1</sup> reported autopsy studies on a patient with a clinical diagnosis of GBS and who had died that showed severe axonal degeneration in nerve roots and distal nerves without evidence of demyelination. It was suggested that this might represent a variant of GBS characterised by an acute axonal neuropathy.

In 1995, Griffin, Ho et al reported on their findings after investigating the yearly epidemic of GBS amongst children in northern China<sup>2</sup>. Twelve autopsied cases were studied. Three of the twelve cases showed the same characteristics of classic demyelinating GBS (AIDP). Six cases showed predominantly axonal damage with only minimum demyelination. (Paradoxically, the other three cases showed only mild changes to the nerve roots and sciatic nerves.) Within the group of six that showed axonal damage, three showed damage to both motor and sensory nerves and three had damage almost exclusively to the motor nerves. The patterns were described as acute motor-sensory axonal neuropathy (AMSAN) and acute motor axonal neuropathy (AMAN).

Of 129 Chinese patients who were studied, 65% had the axonal form, 24% the demyelinating form and 11% could not be classified. One batch of 38 patients (55% axonal, 32% demyelinating, 13% unclassified) was tested for antibodies to the bacterium *Campylobacter jejuni*. Sixty-six percent of the 38 showed evidence of recent *Campylobacter jejuni* infection compared with 16% in the control).

It did not take long for the axonal neuropathy as described by Feasby et al and the 'Chinese paralytic syndrome' to be regarded as one and the same

and it was quickly recognised that *Campylobacter jejuni* was probably the most common trigger for GBS in the West as it seemed to be in China<sup>3</sup>.

In 1997, Ho et al reported<sup>4</sup> on the mechanisms of paralysis and recovery during AMAN. The most severe cases showed degradation of motor axons affecting the ventral roots as well as the peripheral nerves. In contrast, a patient with the characteristic findings of AMAN recovered quickly after plasmapheresis. A sural nerve biopsy proved normal but a biopsy at a neuromuscular junction showed denervation (possibly explaining the Chinese paradox). Antibodies have also been found to be binding to the nodes of Ranvier (between the myelin segments) preventing transmission. There are clearly different mechanisms at work here: one resulting in a slow and incomplete recovery and another resulting in a rapid recovery.

Note: Chinese AMAN patients had been found to recover at an identical rate as Chinese AIDP patients suggesting they fell into the latter category.

So while some patients with 'axonal GBS' may recover quickly, others have considerable axonal damage. They will be joined by those who have bystander axonal damage as a result of AIDP (and indeed CIDP). A problem arises because while demyelination appears to be effectively and promptly repaired by remyelination, axonal degeneration can cause severe persistent disability<sup>5</sup>.

For those left with severe residual effects see our guide *Living with a long term condition*

**Miller Fisher syndrome (MFS)** is also known as:

- The Miller Fisher variant [of GBS]
- Fisher or Fisher's syndrome
- acute idiopathic ophthalmologic neuropathy
- syndrome of ophthalmoplegia, ataxia and are flexia

Related conditions are:

- GBS with ophthalmoplegia
- Bickerstaff's brainstem encephalopathy
- acute ophthalmoparesis

In 1956, Charles Miller Fisher, a Canadian whose specialisation was stroke, described three patients with acute external ophthalmoplegia (eye paralysis), sluggish pupil reflexes, ataxia (lack of balance) and areflexia (absent tendon reflexes). Two patients had no weakness; the other had a facial palsy and possible weakness. All three recovered spontaneously.

Because some patients with GBS had ophthalmoplegia and there were other similarities, Dr Fisher concluded that these patients had suffered a disorder akin to GBS<sup>6</sup>. Pure Miller Fisher syndrome (without generalised weakness) is rare. Electrodiagnostic abnormalities found in all patients are characteristic of an axonal neuropathy or a neuronopathy<sup>7</sup> (predominant sensory nerve changes in the limbs and motor damage in the cranial nerves). Patients described as having Miller Fisher syndrome often have a neuropathy that overlaps with GBS and demonstrate generalised weakness, sometimes paralysis, as additional symptoms. It was sometimes proposed the Miller Fisher syndrome was caused by an inflammation of the brain called a brainstem encephalitis. It is true that the syndrome can be mimicked by a brainstem lesion, but typical cases of Miller Fisher syndrome rarely show any evidence of brainstem abnormalities either radiologically or during post-mortem examination. When clinical or MRI brainstem abnormalities are found, the condition may be referred to as Bickerstaff's syndrome or Bickerstaff's brainstem encephalopathy (or encephalitis) (BBE).

Research in recent years has concentrated in identifying the antibodies that are thought to be responsible for GBS etc. It has been confirmed clinically that MFS, GBS with ophthalmoplegia, BBE, and another condition called acute ophthalmoparesis\* are closely related, forming a continuous range. This is supported by immune findings with the antibody anti-GQ1b IgG being the common factor<sup>8</sup>. This antibody is not found in other GBS patients so it is thought that it is responsible for the ophthalmoplegia. \*Acute ophthalmoparesis (AO) is characterised by acute onset of paresis of the extraocular muscles without ataxia or areflexia.

It has been further noted that many BBE patients have limb weakness and this is considered as an overlap with axonal GBS indicating the disorders are related<sup>9</sup>.

Although the efficacy has not been clinically proven, treatment of Miller Fisher syndrome is much the same as 'classic' GBS though the different

symptoms require modified management with emphasis on the eyes. Intravenous immunoglobulin or plasma exchange treatment is likely in all but the mildest cases. The chances of recovery are good.

### **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 the fluid or plasma. Blood is removed and spun (a bit like the spin cycle of a washing machine) and the cells are separated from plasma. 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 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 very promising 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 & associated chronic variants*.

### **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.

## **What happens next**

### **Going home**

Leaving hospital or a rehabilitation centre and heading home can be daunting and take a while to arrange. You may need equipment to help with everyday tasks, your home may need adaptations or you may need a care package in place to help. There are many people and organisations that can help with this starting with the occupational therapists and your care team. Your family can also be a great help in getting the information together and speaking to organisations that have in depth knowledge of what help is available.

### **Assessment & care plan**

If you are likely to have ongoing health and social care needs you may have an assessment carried out by a multidisciplinary team of health or social care professionals such as social worker, physiotherapists, occupational therapist, psychologists or dieticians. You, and/or a family member should be involved in this process. A care plan should include details of

- the treatment and support you will get
- who will provide support
- when and how often you will get support
- how the support will be monitored and reviewed
- a named person who will coordinate the care plan
- Who to contact

The type of support that might be in a care plan

- Community care services to allow you to live in your home
- NHS continuing healthcare
- NHS funded nursing care
- Rehabilitation or palliative care
- Equipment
- Support from voluntary agencies

### **Residual symptoms**

It is normal to complain of persistent symptoms for weeks and sometimes months after you have been discharged from hospital with GBS. These symptoms vary enormously from patient to patient and include weakness, tingling, and painful tingling, aching in the limbs, cramps and tiredness. It is normal for these symptoms to fluctuate a bit, being worse when you are tired, stressed or affected by an intercurrent illness, such as a cold, sore throat or flu. They gradually wear off, but you may feel some of them coming back in a milder form at times of stresses like that for a year or two. This does not usually mean that the GBS is coming back as recurrence is very rare indeed.

### **Preventative measures**

There is nothing which can be done to alter the very, very small risk of recurrence of GBS. Since GBS occurs after infections, you might think it desirable to avoid contact with infections. To try to do so would be a practical impossibility, and anyway, not worthwhile since you will now be immune to whatever infection triggered your first attack of GBS.

### **Hospital follow-up**

There is a wide range of different practice regarding hospital follow-up visits. Unless you are taking medicines on account of intercurrent medical illnesses, or unless you are being prescribed medicines for pain or complications, you do not need to attend for prescriptions. In fact the person you are likely to need to see more than anyone else is your physiotherapist, rather than a neurologist. However, if you have been very ill and are recovering from a frightening experience such as GBS, it is appropriate to continue to consult a neurologist at appropriate intervals (gradually decreasing) until your health is back to normal, or you have learned to cope with whatever disability the illness has left you with.

## **Hygiene and cleanliness**

Personal cleanliness for those who are unable to attend themselves fully can be a problem. Many returning home from hospital may have reduced use of their hands, usually temporary, but occasionally permanently. Many will be unable to wash themselves, brush their hair, use the lavatory, wipe their bottoms, brush their teeth, cut their nails etc. It is important for both hygiene and self-esteem that these matters are attended to.

## **Teeth**

Through no fault of their own, many people's teeth are neglected during periods of serious illness. Once you have returned home from hospital, arrange an appointment with your dentist as soon as possible. There may be physical barriers making this difficult, as many surgeries have inadequate access for wheelchairs etc. If this is the case there may be a community dental service available that can help. Using an electric tooth brush can be helpful if you have residual weakness in your hands.

British Society for Disability and Oral Health [www.bsdh.org.uk](http://www.bsdh.org.uk)

## **Diet**

During illness, nutritional needs are at their peak, but it is not unusual for patients to lose their appetites or taste for food. Worry and fear often accompany illness and can also contribute to loss of appetite. Good nutrition can be a powerful ally in the process of recovery, ask to speak to a nutritionist for more advice on diet. If taste has been affected, this will usually improve with time. Plastic utensils can be used if bitter or metallic tastes are experienced whilst eating. Sometimes taste changes can be related to medications, but drugs should not be discontinued without first consulting your GP.

## **Exercise**

During the recovery stages, physiotherapy, occupational therapy (OT) and speech and language therapy play a vital role in the rehabilitation process as well as maximizing functional ability. At some point during rehabilitation the rate of recovery will plateau and it is often at this point that patients will be discharged from all the support services on which they may have relied. It is also possible that patients may be placed 'on review'. This means that you

may be followed up at regular intervals and can telephone for advice in-between but don't attend the clinic as often as you did before.

The role of exercise in the ongoing rehabilitation for patients with GBS is still to some extent unclear and clinical trials are being carried out to improve our understanding. However, there is already some evidence that where weakness and fatigue are problems, participation in regular graded exercise can be beneficial.

Exercise can help to improve your muscle strength and reduce your overall sense of fatigue. There are also general benefits of exercise in boosting the immune system, helping your heart and lungs remain healthy and making you feel better about yourself. However, it may take weeks or even months before you feel the benefit of exercise so it is important to pace yourself. Therefore, you should be encouraged to seek advice on whether and how to start regular exercise.

## **Pain**

Pain may never be a problem but can occur in the early recovery phase, The problem does tend to resolve as recovery proceeds.

As pain can make one irritable and difficult to live with at times, it is important that family and friends are kept informed, so that they can understand the reason for such behaviour.

Remember that because the nerves to the hands and feet are the longest in the body, pain will linger in the extremities after it has left other parts of the body.

Pain Concern [www.painconcern.org.uk](http://www.painconcern.org.uk)

Pain Society [www.britishpainsociety.org](http://www.britishpainsociety.org)

Welsh Pain Society [www.welshpainsociety.org.uk](http://www.welshpainsociety.org.uk)

Pain Relief Foundation [www.painrelieffoundation.org.uk](http://www.painrelieffoundation.org.uk)

## **Benefits**

You could be entitled to benefits to help you support yourself.

The benefits system is complex and changes often. Citizen's Advice website has up-to-date information and can help you make an application: <https://www.citizensadvice.org.uk/>

To find out what benefits you (and your family) may be entitled to, you can complete an anonymous benefit check on the Citizen's Advice website or visit your local office and talk to one of their benefits experts.

### **Personal Independence Payment (Pip)**

- For people aged 16 – 64 who have had a disability or long-term health condition for at least three months, which is likely to continue for at least nine months after your claim.
- Points system assesses how your condition affects your ability to cope with daily life and mobility.
- If awarded, there is a daily living component and a mobility component. Each has two rates; standard and enhanced.
- Not affected by income or savings, not taxable and you can get it whether in work or not.

### **Attendance Allowance (Aa)**

- For people aged 65 and over who have a health condition which has lasted at least six months.
- Entitlement is based on care needs resulting from how your health affects your everyday life.
- Not affected by any income or savings you have; payable alongside any other benefits (except Disability Living Allowance or Personal Independence Payments). You do not have to have paid national insurance contributions.

### **Disability Living Allowance (Dla)**

- For adults aged 16 to 64
- Starting from April 2013, this benefit is now being replaced by Personal Independence Payments (PIP). All new claimants must now apply for PIP.
- People who are already in receipt of DLA will be invited to apply for PIP. To find out more use the PIP checker: [www.gov.uk/pip-checker](http://www.gov.uk/pip-checker)
- Some people who currently qualify for DLA will not qualify for PIP, and some who do not qualify for DLA will be able to qualify for PIP.

### **Disability Living Allowance for Children**

- For children aged under 16 who have a health condition or a disability and need help with personal care/supervision or help with getting around outdoors because of this.
- Designed to meet additional expenses of having a child with a long term condition (eg, higher heating bills, special diets, taxi fares, etc).

- Some adults may also still be getting DLA if they claimed before 10 June 2013 but will be invited to claim Personal Independence Payment (PIP). To find out when this will affect you, use the PIP checker: [www.gov.uk/pip-checker](http://www.gov.uk/pip-checker)

### **Employment and Support Allowance (Esa)**

- Payable to people unable to work because of ill health or disability.
- Requires a medical certificate ('fit note') from your GP to make a claim.
- You will be required to fill out a medical questionnaire, attend a medical assessment and a work-focused interview.
- This is intended to determine your capability to work. It is possible to challenge the decision.

**[www.gov.uk/employment-support-allowance](http://www.gov.uk/employment-support-allowance)**

### **Wellbeing**

#### **Sexual relationships**

GBS, CIDP and associated inflammatory neuropathies can bring on problems in any relationship, sexual relationships are not excluded. Dealing with a long term illness or disability can put a great strain on a relationship, particularly when one partner is partially or totally dependent on the other. Even without the actual physical disability, the emotional upheaval can interfere with a couple's sex life and this can be difficult to talk about. This can mean that the once close, intimate relationship can become distant and stressful for both partners. Help is available so speak to your GP or a relationship counsellor.

Other sources or support:

Relate

[www.relate.org.uk](http://www.relate.org.uk)

Sexual Dysfunction Association

[www.impotence.org.uk](http://www.impotence.org.uk)

#### **Emotional issues**

With all the changes in your health it is not uncommon to feel anxious or angry which can be helpful in giving the mental and physical energy that is needed to anticipate and tackle problems. An acute stress reaction is recognised as being an entirely normal part of the process of adjusting to a life change. You can help by seeking information and discussing issues that are worrying you. Most people will make a good psychological recovery but some will continue to experience anxiety and low mood making everyday life difficult.

There is help available and you can speak to your GP or neurologist about

this. They will be able to arrange suitable help for you.

### Other areas of support

Well being [www.nhs.uk/Conditions/stress-anxiety-depression/.../improve-mental-wellbeing.aspx](http://www.nhs.uk/Conditions/stress-anxiety-depression/.../improve-mental-wellbeing.aspx)

Mindfulness [www.bemindful.co.uk](http://www.bemindful.co.uk)

NHS [www.nhs.uk/Conditions/stress-anxiety-depression](http://www.nhs.uk/Conditions/stress-anxiety-depression)

MIND [www.mind.org.uk](http://www.mind.org.uk)

## **Immunisation**

### **What should patients who have had GBS be advised about future immunisation?**

The belief that GBS is an autoimmune condition and the knowledge that immunizations are designed to activate the immune system give rise to continued unease about immunization following the disease. However, many patients have received immunizations after the acute phase of their disease, sometimes repeatedly, without suffering a relapse.

Although there has been concern that some immunisations might have precipitated GBS, there is no hard evidence to support this notion with immunisations which are in common usage in the UK today. However, it would seem unwise for someone whose GBS had come on within six weeks of an immunisation to receive the same immunisation again. Furthermore, many neurologists advise patients not to have immunisation for a year after the onset of their GBS, just in case.

### **Conclusion**

The decision whether to receive a vaccine is an individual one which has to be decided on a case by case basis. Advice should be sought from your own doctor or neurologist.

### **Advice for the carer**

Here are a few practical steps that can help to counteract the stresses and strains of caring for someone suffering from GBS and associated inflammatory neuropathies:

- Gather support from family and friends. Invite help from the local social services/social work department both practical and financial

- Contact a local caring organisation providing support services in your area. They will often help to bath and dress patient, providing a respite so that the carer can go shopping or have a bit of time of their own.
- Take a rest from your duties and allow yourself some personal space. Go for a walk, listen to relaxing music, visit friends etc. Generally take care of yourself, eat healthily, and get plenty of sleep. When friends or relatives visit the patient, take this as an opportunity to have a break and use this time to do something for you.
- Take the pressure off by putting some activities on hold.
- Be mindful of the patient's limitations.
- Talking is therapy and you may also find it useful to speak to an external source: friends, relatives, caring organisations, **GAIN** etc.
- Get organized. Investigate benefit entitlements with the hospital social worker and/or Social Services/Social Work Department. Liaise with the hospital occupational therapist (OT) and physiotherapist about equipment arrangements.
- Be temperature conscious if the patient is suffering from lack of sensation, ie run and test the bath water. This also applies to the cooker, iron etc. as there may be no sensation and a patient can get burnt or scalded very easily.
- Taste buds may be affected for a while, so prepare meals to suit the patient. Vitamin supplements can be included if a balanced diet cannot be achieved.
- Beware of falls brought about by weakness or unsteadiness.
- Be mindful of potential accidents resulting from weakness and/or numbness (ie dropping things). Care should be taken when the patient is using hot appliances, such as when cooking or ironing. A microwave oven is a very convenient, safe way of preparing food.
- Help the patient with daily exercises. Ensure that everything is done in moderation and that the patient does not start rushing around too soon. Encourage the patient to talk openly about his/her experiences and fears.

Carers UK

[www.carersuk.org](http://www.carersuk.org)

Carers Scotland

[www.carerscotland.org](http://www.carerscotland.org)

Carers Northern Ireland

[www.carersni.org](http://www.carersni.org)

Carers Wales

[www.carerswales.org](http://www.carerswales.org)

The Carers Association ROI

[www.carersireland.com](http://www.carersireland.com)

### **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 to six weeks although it typically happens within a few days.

The initial progressive phase lasts longer, and usually it is much longer, than 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.

If you wish to know more about CIDP, see our guide titled *CIDP & associated chronic variants*.

### **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.

- <sup>1</sup> Brain 1986 Dec;109 (Pt 6):1115-26
- <sup>2</sup> Brain 1995 Jun;118 (Pt 3):577-95, 597-605
- <sup>3</sup> Hughes RA, Rees JH, Infect Dis 1997 Dec;176 Suppl 2:S92-8
- <sup>4</sup> Neurology 1997 Mar;48(3):717-24
- \_\_\_\_\_
- <sup>5</sup> Hughes et al, Mult Scler 1997 Apr;3(2):88-92
- <sup>6</sup> Fisher CM. Syndrome of ophthalmoplegia, ataxia and areflexia. N Engl J Med 1956;255:57-65
- <sup>7</sup> Fross RD, Daube JR. Neuropathy in the Miller Fisher syndrome: clinical and electrophysiologic findings. Neurology 1987 Sep;37(9):1493-1498
  
- <sup>8</sup> J Neurol Neurosurg Psychiatry 2001 Jan;70(1):50-55
- <sup>9</sup> Yuki, Rinsho Shinkeigaku 2004 Nov;44(11):802-4

## Have you been diagnosed with one of these conditions?

### Acute

**GBS** Guillain-Barré syndrome

**AIDP** Acute Inflammatory Demyelinating Poly(radiculo)neuropathy

**MFS** Miller Fisher syndrome

**AMAN** Acute Motor Axonal Neuropathy also known as Axonal GBS

**AMSAN** Acute Motor Sensory Axonal Neuropathy

### Sub-acute

**SIDP** Sub-acute Inflammatory Demyelinating Poly(radiculo)neurathy

### Chronic

**CIDP** Chronic Inflammatory Demyelinating Poly(radiculo)neuropathy

**CIAN** Chronic Idiopathic Axonal Neuropathy

**CMFS** Chronic Miller Fisher syndrome

**CANOMAD** Chronic Ataxic Neuropathy with Ophthalmoplegia, IgM Monoclonal gammopathy, cold Agglutinins and Disialogangliosides antibodies

**MMN** Multifocal Motor Neuropathy

**MMNCB** Multifocal Motor Neuropathy with conduction block

**MADSAM** Multifocal Acquired Demyelinating Sensory and Motor neuropathy also known as Lewis-Sumner syndrome

**PDN** Paraproteinaemic Demyelinating Neuropathy sometimes described as CIDP with Paraproteinemia

**POEMS** Polyneuropathy, Organomegaly, Edema/Endocrinopathy, Monoclonal gammopathy and Skin changes syndrome

**Contact Guillain-Barré & Associated Inflammatory Neuropathies for more information**

This guide has been written by the GAIN Medical Advisory Board and gives an honest account of the conditions. Not all content will apply to you and if you need more information ask your consultant or GP.

To find out what other help we can provide please contact us or visit our website.

**Guillain-Barré & Associated Inflammatory Neuropathies (GAIN)**

Woodholme House  
Heckington Business Park  
Station Road  
Heckington,  
Sleaford  
Lincolnshire, NG34 9JH



+44 (0) 1529 469910



office@gaincharity.org.uk



Monday to Friday 9am – 3pm

[www.gaincharity.org.uk](http://www.gaincharity.org.uk)

Registered charity 1154843 & SCO39900