

# GUIDE

## *Severe GBS*

This series of guides is produced by the Guillain-Barré Syndrome Support Group. We are a registered charity that supports those affected by the Guillain-Barré syndrome (GBS) and related conditions in the United Kingdom and the Republic of Ireland. The related conditions include chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) and Miller Fisher syndrome (MFS).

Our guides are easily downloaded from our Web site at [www.gbs.org.uk](http://www.gbs.org.uk) in PDF format and may be both read and printed using free Adobe Reader software. Alternatively, you can request printed copies from our office.

**For information and support, ring our helpline on 0800 374 803**

In the Republic of Ireland, call 0044 1529 415278

### **Definition**

‘Severe GBS’ is regarded in this document as GBS that results in severe and permanent disability. In many cases, the affected person will require some ongoing care from others.

### **Introduction**

This supplement to our publication *After GBS* has been written by a person who has had severe GBS. The text has been checked for medical accuracy by a neurologist and approved by the Support Group’s Medical Advisory Board. It should be appreciated that this supplement only applies to a small percentage of GBS patients. Even if a patient appears to be very ill in the acute stages of GBS, it does not follow that the patient has ‘severe GBS’ as defined above. We recommend that if you are seeking information about a patient acutely ill in intensive care, then you should read our booklet *The GBS Patient in Intensive Care* in the first instance.

Since treatment with plasma exchange in the 1980s and that of intravenous immunoglobulin in the 1990s, the average time taken for patients to recover from GBS has reduced. Unfortunately,

the percentage of patients that are left with severe disability has only fallen a little and is still about 5 - 10%.

The reason for this paradox is perhaps because in some patients, the illness damages the peripheral nerve cores or 'axons' severely whereas the insulation or 'myelin' is the major target in most patients. Though the treatments reduce myelin damage and so allow faster recovery, they do not protect the axons from damage so effectively.

An axon is the long fine strand of a single nerve cell or 'neuron', the nucleus of which is in the dorsal root ganglion, spinal cord or brain. If that cell dies, then there is no conduction path for nerve signals. In severe GBS, the axons are so severely damaged that they are severed at the point where they are inflamed. The distal part (the part on the side away from the centre) regrows from the centre outwards. The rate of growth is estimated as 1 mm per day. Since the longest nerves in the human body are around one metre long, it would be expected to take almost three years for the longest nerves to regenerate if they were divided near their cell bodies. In practice, the regrowth and recovery are often incomplete.

Most recovery takes place in the first year after which it tends to slow down. Some recovery continues for several years but after three years, recovery is exceedingly slow and usually minor. However, minor improvements may make a considerable difference to quality of life. They can therefore be of some significance.

## **Anxious times**

Patients and their relatives and friends will have been given a good idea of what to expect in the recovery phase of the illness. In most cases, these expectations are borne out and a steady improvement will be noticed. But sometimes improvement is just too slow and it becomes obvious that recovery will not happen within a few months. There are some rough pointers as to what may happen. Elderly patients with an explosive onset in the acute stage and those who need ventilation, especially for an extended period, tend to do less well. But these are only indicators. Some very old patients do very well. Some patients ventilated for long periods do recover completely or almost completely. Some patients who are not ventilated are left with significant disability.

If recovery remains slow, doctors may order some electrical tests to ascertain axonal damage, if they have not already done so. If the phrase 'axonal damage' is mentioned, then it would certainly be worth asking for an interview with the consultant neurologist to discuss the possible implications.

## **Specialised treatment**

With the introduction of intravenous immunoglobulin (IVIg) treatment, it is more likely for GBS to be treated at a local general hospital rather than transferring the patient to a specialised neurological centre. Provided the intensive care facilities at the local hospital are adequate, the advantages of being treated close to home probably outweigh the disadvantages. If a patient needs prolonged ventilation or there is doubt about the diagnosis consideration should be given to being transferred to a neurological centre.

## **Prognosis**

The prognosis varies enormously. GBS can prove fatal though this is usually because of breathing or heart complications or another coexisting condition. Some become so severely paralysed that the ability to breathe never returns; others require ventilation for many months, even a year or more, before they can be weaned off the machines. These patients are often confined to wheelchairs and only have the rudimentary use of their arms and hands. Such very severe cases are fortunately very rare.

At the other end of the scale are those patients who make a full recovery. Indeed, most patients do make a full recovery but there is always going to be a percentage who fall between the two extremes. Trying to categorise them all is an impossible task as different people have to cope with different residual problems. Those with minor disability invariably have problems with their hands or feet. As the degree of disability increases, then problems in the limbs increase and arms and legs are also affected.

The autonomic nerves, ie those that control the heart, bladder and bowel function, temperature control and sexual function, may be affected by GBS. As function returns to the rest of the body, then the autonomic nervous system recovers too. Persistent problems affecting the bladder and sexual function sometimes occur.

## **Coming to terms**

Depending on the degree of residual disability encountered, consequences can range from being relatively minor to being completely devastating. The former cannot be belittled as they might, for example, necessitate a change in profession, though this is in a different league when compared with a person who is left effectively quadriplegic requiring a very high degree of personal care.

The doctor of a patient with a spinal injury can often make a relatively rapid prognosis and, if bad, can break it to the patient there and then. These are desperate times but at least under these circumstances, patients and their families start to begin to understand the consequences of such information. For those with potentially severe GBS, things are far more uncertain and the consequences can be very stressful. Well-meaning people frequently tell patients, even those profoundly ill, that they will get better. Doctors are usually more reserved and will substitute 'should' for 'will'. But when recovery does not take place as predicted, very little information is sometimes offered, leaving patients and families to figure the consequences out for themselves.

## **Immediate actions**

Anyone who has been in this situation will agree that the greatest fear is the fear of not knowing. You should seek a consultation with the consultant neurologist and express these fears. You are also welcome to contact the GBS Support Group. If you call our helpline, remember that the volunteer answering your call may not be very familiar with severe GBS and the same might apply to a local volunteer who might contact you. If necessary, ask for one of our specialist severe GBS contact persons to get in touch. Alternatively, a call or e-mail to our office will get you in touch with an experienced volunteer quickly.

Of prime consideration is the patient's morale. Though the outlook is uncertain, being too

optimistic now may lead to problems later. On the other hand, being too pessimistic will ruin morale that might later prove unfounded. Equally important is the morale of the family members. Carers must put themselves first because if they do not look after themselves, they cannot possibly care for others.

Ensure that the patient is physically comfortable. If the patient is in pain, insist it is treated by a specialist in nerve pain. If the patient complains of aches due to lack of turning, insist that it is carried out more regularly. If the patient is in a ward where all hope of dignity has long since disappeared and where he or she has to fit into an almost military regime (eg bowels by numbers), perhaps it is time to move on to a different environment.

Now is the time to consider benefits and allowances. Because of constantly changing rules, this supplement cannot be too specific but the hospital multidisciplinary rehabilitation team will have the information and forms to hand. A problem with GBS is that it usually does improve so it can be difficult to convince the adjudicators that the disability is not short term. A covering letter from the consultant accompanying any application is a good idea. Some patients will oppose obtaining benefits because in so doing, they are admitting that they are not getting better. Explain that the benefit may be temporary and in any event, it is money that has been earned. Delaying application can lose hundreds of pounds as awards are not made retrospectively. If an application fails, you must appeal at once. Contact the Group for further advice.

## **On to rehabilitation**

It is a matter of concern, that in the twenty-first century the Group still receives stories concerning the thoughtless treatment of some patients. Thankfully, stories of praise are far more common but this does not diminish the unnecessary anguish suffered by a minority. Most problems stem from a lack of finance or plain ignorance. The slow recovery often associated with GBS sometimes leads to premature discharge on the grounds of 'lack of progress'. Other examples include patients being transferred for rehabilitation to units specialising in inappropriate disciplines.

If a patient is being transferred to a rehabilitation unit seek reassurance that it is one which is appropriate for people with purely physical problems and not one which is designed for brain-damaged patients or even for people with spinal injuries. Their needs are quite different from those of patients with the after-effects of GBS which may make it difficult for the GBS patient's special needs to be understood, especially by junior members of the rehabilitation team.

Another problem arises when poorly informed staff compare one GBS patient with another. A patient who fails to progress as another has progressed is liable to be branded as lazy and uncooperative. The ignorance of the possibility of severe GBS and its slow recovery can be a cause of considerable anguish.

Perhaps too much is expected of rehabilitation. If a patient is making slow progress, a transfer to a rehabilitation unit will not speed it along. Indeed, a regime of intensive therapy is as likely to do as much harm as good. Ideally, once able to leave the acute hospital, the patient should be admitted to a specialist neuro-rehabilitation unit that understands the problem. It could be that the patient may take years to maximise potential. It would be asking too much of the unit to expect a patient to stay that long and it would be bad for the patient too. But the patient should be given the time to obtain a high degree of the expected potential. This will make the

eventual return to the home environment less challenging as the patient will have obtained valuable extra independence.

## **Discharge**

The consequences of severe illness are rarely discussed. All too often it is assumed that the patient will be discharged home to the care of a spouse, a partner, parents etc. But some severe GBS patients will be profoundly disabled: unable to walk, dress, feed themselves, use a lavatory etc. These patients will need full-time care though the requirement may become less as improvement continues. But suppose a patient has no-one who is prepared to be a carer? What happens if a patient was in a rocky relationship before being taken ill? The chances are that the relationship will have long since ended. Suppose a spouse has to continue working to maintain an income? Parents may be too old or infirm to cope.

This is a very complicated area and no two cases are ever the same. Inevitably in these 'social' cases, the social services will become increasingly involved.

If profoundly disabled, those approaching discharge without anyone to care for them have few options. To the majority, living in an institution would be anathema. The luckier ones have their own homes that after adaptation they could live in, provided a care package could be funded. Those without existing homes have the added burden of finding one.

Inevitably, much of the responsibility falls on local social services and times are not easy. Most local authorities will eventually comply with their statutory duties but it can be a hard fight. Be prepared to write a great many letters and never accept 'no' for an answer. If you receive the reply that what you wish is 'against policy', shame those who made it and get it changed.

A good source of help is RADAR's *Directory for Disabled People*. It is described as a comprehensive handbook of information for everyone involved with disability, a guide to the whole range of services, facilities and opportunities available. It covers statutory services, benefits, aids and equipment, independent living, caring, housing, education and employment, motoring and mobility, holidays, arts, sports and leisure, sex and personal relationships, and access. It also covers relevant legislation, information and legal services and useful contact organisations.

## **The home**

Assuming the discharged patient uses a wheelchair, a bungalow or ground floor apartment is generally superior to any other type of house. Ideally, the home will have amongst other things: level access, doorways widened from the old standard of 2ft 3in to 2ft 6in or more, doors and corridors so located as to ensure access, lowered light switches, a bathroom with a suitable bath or shower, and a lavatory with a seat raised to the height of the wheelchair cushion and plenty of room to manoeuvre. Powered chairs generally need more space than manual ones.

The reality will usually be some kind of compromise. However, it is worth being imaginative and before building an extension to an otherwise unsuitable house, consider a move or even building from scratch. The latter will take time but will make life much easier.

If a patient can self-propel a wheelchair, the chances are that carpets will prove difficult to move over. If possible, replace pile carpets in the most-used rooms and corridors with office-type short pile carpets or consider wood or laminate flooring.

An occupational therapist attached to the local social services will become involved with the case to discuss what needs to be done to make the home environment practical and safe.

## **The care package**

Providing care may not be as daunting as it may at first appear. Multiplying an hourly care rate by the number of hours in a year can produce figures resembling telephone numbers. Clearly for long-term care, a different approach is required. For example, if a working partner can cover the nights and weekends, only the weekday working hours need to be provided for. If there is no partner, night cover is usually available at very reasonable cost. There are plenty of people who work during the days who are willing to provide this cover to make additional income.

Care during the day may not need to be continuous. Many confined to wheelchairs will be quite capable of being left alone for an hour or two, maybe longer, provided they have access to back-up should it be needed.

Your local social services will appoint a social worker to the case who will co-operate closely with the hospital staff. In spite of your fears, a patient will not be discharged without sufficient care being provided, though should you have any cause to question this, you must discuss it.

## **Keeping the mind active**

The patient on returning home will have limited mobility and will have problems to a lesser or greater extent manipulating with hands and fingers. One of the greatest tools that has revolutionised the lot of disabled persons is the personal computer. It is very unusual for a person not to be able to use a computer in one way or another and for most disabled people, computers put them on level terms with everyone else. The advent of the World Wide Web and electronic mail have transformed people's lives and brought about the reality of people returning to valuable employment.

If the patient cannot type there are plenty of alternative ways of producing input. When discharged from hospital, the ability to use fingers or a tool held in the hand might be some way off, so consider having a dental technician making a rubber-tipped stick to hold between the teeth. Controlling a mouse may be difficult so try a trackball. There are alternative keyboards available too, as well as other devices for those who cannot use their hands at all.

If a patient is not interested in computers, remember that the computer is a wonderful tool for contacting others who share your real interests. There are plenty of able and willing people who will sort out the technicalities. New possibilities of on-line services such as shopping and banking are making huge differences.

Books can be more problematical. Many people with poor use of the hands will develop their own techniques for reading books and other publications. If a person cannot turn pages, then there are page-turning devices available. These are fine if reading from one end of a book to another but remain difficult if searching around a textbook for example.

## **Mobility and transport**

A severe GBS patient is likely to be wheelchair-bound for an extended period if not permanently. If the chair can be self-propelled, that is a distinct advantage as it forms a vital part of the exercise regime. If possible, opt for a light chair made from a modern alloy. Such chairs are easier to propel for both user and carer and have infinite advantages when, for example, putting in the back of a car.

Electric scooters may be suitable for some people, but a GBS patient who cannot use a self-propelled wheelchair will most likely need an electric wheelchair with a joystick for control. If their hands and fingers are not usable, then an alternative method of control has to be devised. When obtaining any wheelchair by any means, it is crucial that the best, unbiased advice is sought.

Ensure that any wheelchair is provided with a suitable cushion. Many privately bought chairs will be provided with an adequate though not a top-of-the-range cushion. Other chairs may come with no cushion at all. It should be possible to obtain a suitable cushion on prescription for even a privately obtained chair. Some cushions are inflatable with a pattern of cells like an egg box. More stable are the cushions filled with 'goo' that moulds perfectly to any bottom.

Those who use wheelchairs will be in receipt of the mobility component of the Disabled Living Allowance or equivalent. As such, they can buy or hire a car using the Motorability scheme though some manufacturers offer a discounted scheme that can sometimes prove slightly cheaper. The choice of vehicle and any necessary adaptations, is decided upon by the capabilities of both patient and carer, and whether the patient intends to drive the vehicle.

Public transport is patchy. Even if wheelchairs can be loaded onto buses and trains — and the situation is improving — hanging on for dear life may be the order of the day. Flying is generally easier though disabled passengers sometimes suffer the indignity of being carried up the steps. Passengers requiring assistance are inevitably the first to be boarded and always the last off so early check-in is essential. Plenty of time must be allowed for connections. Airlines must be informed of needs when booking. Perhaps the biggest potential problem whilst flying is using the toilets. The best advice may be to break a long flight up into two or more sectors. Steps should be taken to reduce problems with bowels. Long drinks should be avoided before flying. Because flying is dehydrating, drink is essential. Drink only moderately but often. Alcohol should not be taken.

## **Personal problems**

Anyone reading this supplement will be quite familiar with the problems of bladders and bowels and how they are dealt with. Fortunately, most GBS sufferers, even those with severe GBS, do regain a near normal bladder and bowel function. Constipation and urine retention are more common than incontinence. So problems tend to be incidental like getting on and off a WC or, in the case of a man, using a bottle while seated in a chair.

Hardly ever discussed in hospital is the subject of sex. Every bodily system is monitored and checked save this one. The problems are various and differ from person to person and male to female.

In men, impotence or partial impotence is common, due to damage to the autonomic nerves in the pelvic region. This may be treatable but if there is additional weakness, it may be necessary

for a female partner to take a more dominant role! The only advice the author (male) ever received on the matter was from a very attractive female doctor: 'You'll have to be adventurous.' Anecdotally, more females than males suffer from a lack of libido, sometimes due to profound discomfort. Whether the victim is male or female, his/her partner has to demonstrate a great deal of understanding.

At the other extreme there are those who are profoundly disabled, still have sexual urges but no partner. Though this is a very delicate subject, it is not going to disappear by pretending that it does not exist.

Women, of course, run the additional possibility of becoming pregnant. Though pregnancy and GBS are not mutually exclusive, such an outcome could further complicate an already complicated situation.

The Support Group has published a leaflet on sexual relationships after GBS and is useful additional reading. If you have not received a copy with this supplement, please contact our office for a copy or visit our Web site at [www.gbs.org.uk](http://www.gbs.org.uk).

If after reading this guide you still have anxieties and unanswered questions, telephone our helpline on 0800 374803 (UK) or 0033 1529 415278 (RoI). Alternatively, you can e-mail us or register for support on-line

The GBS Support Group is a registered charity and receives neither government nor Lottery funding. If you have found this guide helpful and would like to help us to continue publishing copies for others affected by GBS and its related conditions, please consider making a donation to the Support Group. Secure donations may be made on line. Alternatively you can request a form from our office.

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