

GBS

Guillain-Barré Syndrome Support Group
of the United Kingdom Registered Charity 327314

Childhood Guillain-Barré Syndrome

A guide for parents and carers

First Edition 1998

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Information

Publications

This booklet has been produced by the GBS Support Group of the UK, a registered charity founded in 1985. The booklets available from the Group are:

- *Guillain-Barré Syndrome* — a guide for patients, relatives and friends;
- *The Guillain-Barré Syndrome Patient in Intensive Care* — a guide for relatives and friends;
- *Childhood Guillain-Barré Syndrome* — a guide for parents and carers;
- *CIDP* — a guide for patients, relatives and friends; and
- *Jenny's Story* (a beautifully illustrated booklet for children with GBS).

The Group's address is: GBS Support Group of the UK, LCC Offices, Eastgate, Sleaford, Lincs, NG34 7EB. Tel/fax: 01529 304615. E-mail: admin@gbs.org.uk. Under certain circumstances, copies may also be obtained by calling the Free Helpline (see below). If you live outside the UK or Irish Republic, see page ii.

Free Helpline

If you are presently suffering from GBS or a related condition, or are a relative or friend of a sufferer, and wish to talk to someone now about the illness, call the Free GBS Helpline on **0800 374 803** (UK only). Our Helpline volunteers are themselves former patients or their carers who are able to understand your anxieties and answer your questions.

Please do not use the Helpline for routine or administrative calls. Requests for publications by others than patients, relatives or friends should be made to the GBS Office.

Hospital visits — local and specialist contacts

Nothing can boost the morale of a GBS sufferer in hospital more than a visit by a recovered patient. The Group has a network of over 100 local and hospital contacts who carry out this important service and there is probably one in your area. Most importantly, there exists someone locally who understands the sense of isolation that affects all those who come into contact with GBS, and who can offer practical help.

There are occasions when local contacts do not have the necessary knowledge for particular cases (eg childhood GBS, CIDP, severe GBS). Under these circumstances,

the help of one of our specialist contacts can be called upon.

The Group's Help Fund

The Group's Help Fund exists to provide emergency financial support at the most critical of times. The fund has not been established to pay for care and equipment, provision of which are the responsibilities of the statutory services, nor can it make ongoing grants to supplement statutory benefits.

An example of how the Fund may help is the case of a relative having difficulty affording the costs of visiting a GBS patient, especially if the patient is in a hospital many miles from home.

Requests to the Fund should be made in writing to the Office and be supported by a covering letter from a health professional.

Cost

The above information and services are provided absolutely free. There is no obligation to join the Support Group though many who register with us do subsequently become members.

Registration

If you are a sufferer, or the relative or friend of a sufferer, and have received this booklet directly from the Support Group, then you will be already registered with the Group. Those who are registered receive free copies of the Group's journal *Reaching Out* for a year. If you received this booklet in some other way (eg from a friend, a hospital or downloaded from the Internet), by registering you too will receive free copies of the Journal.

Our records are kept in the strictest confidence and will not be divulged to third parties. To register with the Group, please fill in the form on page iii.

Membership

If you wish to become a member of the Support Group now, fill in the membership form that accompanied this booklet. If the form is not available, please contact the office for a replacement or visit the Group's Web site.

Other countries

If you live outside the UK or the Republic of Ireland and are seeking support, we will make every effort to put you in touch with a local support group should one exist. Since the Group's information can be readily downloaded from the Group's Web site

or attached to electronic mail, the Group's publications are not normally posted overseas. If receiving the information in this way is impossible or unsatisfactory, a donation to cover the airmail charges would be much appreciated.

The Group regrets that it does not have the resources to register cases from outside the UK or the Republic of Ireland and supply free copies of its journal. However, it is possible to become an 'Overseas Member' of the Group and receive copies of the Group's journal by air mail. Please see the membership form that accompanied this booklet. If the form is not available, contact the office for a replacement or visit the Group's Web site.

Disclaimer

The GBS Support Group of the UK is not a medically qualified organisation. The information it publishes is for general educational purposes only and should neither be regarded as advice on the diagnosis or treatment of the Guillain-Barré syndrome nor any other medical condition. The information provided by the Group is designed to support, not replace, the relationship that exists between a patient and his/her existing doctor. While every effort has been made to ensure the accuracy of the

Registration form (UK/Ireland only)

Please read the paragraph 'Registration' on the previous page before filling in this form.

Please fill in as much detail as possible in the boxes below. Information will be treated with the strictest of confidence. Please add any further details and requests overleaf.

YOUR DETAILS:

Name	<input type="text"/>
Address	<input type="text"/>
Town	<input type="text"/>
County and post code	<input type="text"/>
Tel	<input type="text"/>

PATIENT'S DETAILS:

Name	<input type="text"/>
Home address	<input type="text"/>
Town	<input type="text"/>
County and post code	<input type="text"/>
Tel	<input type="text"/>
Hospital	<input type="text"/>
Ward/unit	<input type="text"/>
Town	<input type="text"/>
Relationship to you	<input type="text"/>
Date entered hospital	<input type="text"/>
Diagnosis (if known)	<input type="text"/>
	GBS/CIDP/other(state)
Ventilated	YES/NO <input type="checkbox"/> <input type="checkbox"/>
	Age <input type="text"/>

If you would like to receive a call from your local GBS contact, who will be pleased to offer support and/or arrange a hospital visit, please tick box.

Send this form to:

**GBS Support Group of the UK
LCC Offices, Eastgate**

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Childhood Guillain-Barré Syndrome

A guide for parents and carers

Introduction

This booklet is designed to help carers of children who have Guillain-Barré syndrome (GBS). By reading this you should have an idea of what is involved with the condition and how it may affect your child. However, it is important to remember that this book is a guide only and every child is different. If there is anything you do not understand, please talk to your doctor. He or she will always do his or her very best to help.

Incidentally, another name you may hear in relation to GBS is **acute inflammatory demyelinating poly[radiculo*]neuropathy** (AIDP). Several other similar terms are occasionally used.

Definition

Guillain-Barré syndrome is an uncommon condition that occurs in people of all ages. However, of all the peripheral neuropathies in children, it is now one of the commonest. It affects around 1 in 100,000 children in the UK every year. It is a condition that affects the nerve sheaths, but does not usually affect the brain or the spinal cord. The nerves become stripped of their outer insulating coat (called myelin) — this means that the nerve cannot conduct messages so quickly and occasionally not at all. We do not completely understand why this happens; it is an area of ongoing research.

*'Radiculo' is sometimes omitted.

However it is generally accepted that GBS is an autoimmune condition most commonly triggered by a passing previous infection which stimulates the body's immune system into forming antibodies which mistakenly attack the myelin nerve sheaths. Eventually the nerves are able to recover.

Clinical course

GBS usually occurs around two weeks after a respiratory (eg a cold or 'flu) or a gastro-intestinal (eg diarrhoea or vomiting) infection. More rarely it may follow other infections such as chicken pox. We do not know why it affects a particular person as more often than not the whole family has had



Doctors will always do their best to help.

the infection but usually only one person will go on to get GBS. Furthermore, the person affected will not necessarily have been the most unwell with the initial illness.

The typical pattern of development is a gradual onset of symmetrical weakness starting in the feet and sometimes hands and slowly spreading upwards. Although the main problem is with weakness, another feature is cramping muscle pain and back ache in many children.

Along with the weakness of the arms and legs, the nerves of the head and neck may be affected producing facial weakness with loss of facial expression. There may be difficulty moving the eyes, swallowing and talking. Speech may take on a nasal quality. Hearing, however, is rarely involved. Weakness may also progress to involve the muscles of breathing requiring extra assistance (see later). Some children have involvement of their autonomic system (blood pressure, heart rhythm, bladder, bowels and temperature control). This complication is less common in children than adults with GBS. Some children have pain in their muscles (usually arms and/or legs) described as shooting or cramping in nature. Unfortunately the pain can be quite uncomfortable, however there are some very good medicines available.

Despite, sometimes, severe weakness and tiredness, your child is conscious and needs to be told what is happening and will be reassured by familiar faces and voices.

You may also notice that your child is more tearful and moody than normal. Although this may be completely appropriate in the current situation, children can continue to have fluctuating mood swings some time after recovering from GBS.

The weakness usually worsens over a one to two week period in most children until it reaches its peak, which may last from a few days up to several weeks. This is called the 'plateau period'. Following this plateau, recovery begins.

Diagnosis

GBS is diagnosed by a combination of features including the above history. On clinical examination the doctor will find evidence of a peripheral neuropathy noting the child's weakness, absent tendon reflexes and sometimes altered sensation in the hands and feet (described as a 'cotton-wool' sensation in a 'glove & stocking' distribution).

To be certain of the diagnosis, the doctor will need to perform a lumbar puncture to measure the protein level in the cerebrospinal fluid (the fluid bathing the spinal cord), which rises in the second week of the illness.



Reduced or absent tendon reflexes may indicate GBS.

This test consists of inserting a small needle into one of the spaces between the vertebrae low down the back just long enough to collect some fluid. The test is carried out on children with them lying on their side and curled up in a ball. The nurse usually puts local anaesthetic (Emla) over the site of the lumbar puncture and the doctor may inject more local anaesthetic before doing the test. In order for the results to be accurate, it is very important to keep as still as possible when the needle is being inserted. The test itself only takes about five minutes to perform but the careful preparation takes longer. Lumbar punctures are frequently performed in children's medicine for all kinds of reasons and children tolerate them very well.

The other important test is to measure the way the nerves conduct messages. This is called a nerve conduction study and takes about 20 minutes to carry out. The test involves stimulating the nerve and this has the sensation of a tapping or jolting. The doctor may also feel that it is necessary to measure the activity in the muscles as well. This includes placing a very small needle into a muscle and taking recordings. It is very quick and only hurts a tiny bit when it first goes in. If you distract your child just before it happens he or she may not even notice.

The doctor is likely to take some blood samples as well; he or she can use the Emla cream again so that it does not hurt.

Differential diagnosis — what else could it be?

Not all children present with the classical history described above. It is very important for everyone to feel confident with the diagnosis of GBS as this would affect the management of your child. Some children can have many signs in common with GBS but not have the condition. Thus the doctor may need to perform a number of other tests to ensure nothing is missed, especially if it may alter the way your child is treated. The investigations are likely to include blood tests, urine samples and some body swabs. It may be necessary to perform a lumbar puncture in the first week of presenting if there is any fear of ongoing infection. Rarely, a 'follow up'

lumbar puncture may be needed a week later. Some children have neuroimaging of their head and/or spine (by a CT or MRI scan) when the diagnosis is in doubt. The doctor will try to keep the tests to a minimum but it is important nothing is missed. Make sure you understand what each test is for, otherwise it may be a big surprise to find your child's treatment suddenly changing.

Management

It is important your child is managed in a centre familiar with GBS and with intensive care facilities, so your child may be moved to a hospital you do not routinely use. Most of your child's care will be 'supportive' for breathing, feeding, bowel or bladder functions. It will also depend on how weak he or she becomes. Physiotherapy is needed to ensure good joint mobility and to keep the chest clear. Splints may be used to keep the hands and feet in a normal comfortable position. Frequent turning is important, if your child is unable to do this for him/herself, to prevent pressure sores. If there is any difficulty in swallowing then a naso-gastric tube (NG) may be passed to ensure adequate nutrition is maintained. Sometimes it is necessary to supply fluids by an intravenous (IV) line. This may occur if your child becomes very tired. A full stomach from NG feeding puts too much pressure on the diaphragm and makes breathing difficult.

Very rarely children find they have problems passing urine or become constipated. If this occurs the urinary symptoms are easily helped by passing a small catheter into the bladder to drain all the urine off. Constipation is managed with medicines and doesn't tend to be a major problem.

Children may become very distressed with muscle cramps or back ache and the doctor will be happy to prescribe painkillers for this. It is however important to remember that it is better to avoid analgesics which are sedative.

During the illness, it is important to keep any monitoring device the doctors and nurses advise on your child. He or she may become irritated with it, but it must remain. Most children will need monitoring of their heart

rhythm (pulse), breathing (oxygen saturation) and blood pressure at regular intervals. The doctor will also be concerned about the breathing and will ask your child to blow into a machine to measure the 'vital capacity' at regular intervals. If your child is too young or weak to do this then the doctors and nurses will watch the breathing pattern carefully to check it does not become too laboured and the doctor may need to do blood tests to monitor it.

Treatment

Steroids

Steroids were used in the past but are only felt to be of use in exceptional circumstances now.

Immunoglobulins

High dose intravenous immunoglobulin (IVIG) has been shown to speed up recovery. The infusions are usually commenced when weakness is so severe that it is not possible to walk or there is evidence of respiratory involvement (difficulty in breathing). Immunoglobulin may be given on the ward or the intensive care unit. A few children are allergic to this treatment and develop a rash with it. If this happens the infusion is stopped. It may be restarted at a lower rate provided the rash has settled completely.

Plasma exchange

Plasma exchange has also been shown to speed up recovery, but it is more complex to administer than IVIG. The child requires several large intravenous lines for this procedure and about 250mls (½ pint) of blood are removed at a time in a closed circuit and 'washed' before re-entering the child. By doing this an attempt is made to remove the circulating antibodies in the blood which are attacking the nerves. This would only be performed in an intensive care unit and although it is quite an invasive treatment, it is frequently performed in the above setting for other conditions. The exchange may need to be performed on several occasions until evidence of improvement is seen.

Paediatric intensive care

Around 10% of children with GBS will become so weak that they cannot breath without the support of a ventilator. Understandably this can be a frightening situation for a child who is still fully aware of everything going on around him or her. Parents and carers must provide the child with all the positive support needed to avoid unnecessary trauma.

Things you can do to help

A calm atmosphere is essential. Remember, everything you say in front of your child is likely to be heard. So it is important to be reassuring. Talk about things that matter to your child (a pet, the football results, family events). Bring a favourite toy. Your child may become extremely frustrated especially if he or she cannot speak. Try to work out ways of communicating. The hospital staff should be able to provide you with aids on how to do this (eg picture cards for eye pointing). The doctors and nurses are able to monitor evidence of your child becoming distressed by readings of the pulse, blood pressure and rate of breathing: so called 'vital signs'. The best help will be a close member of the family providing reassurance, however light sedation will be used whenever necessary. During the time on the intensive care unit, support will continue with physiotherapy, IV fluids and NG feeds as before. The length of time children require artificial ventilation varies. Many show an improvement within as little as a week. The intensive care doctors will try therefore withdrawing the ventilation after this time. If your child is still too weak, the doctors may arrange for a tracheostomy tube to be fitted which will allow ventilatory support to be continued with less discomfort for as long as necessary. The tracheostomy tube is inserted into a small hole made in the front of the neck during an operation. To this tube the ventilator tubing can be connected instead of to a tube in your child's nose or mouth. When your child can breathe without support, the tracheostomy tube can be removed and the hole will close on its own.

Outcome/rehabilitation

The majority of children with GBS make a full recovery without any signs of having had the condition. However a small number may have some persisting problems. The commonest complaint is weakness of the hand and foot muscles. Most recovery is seen in the first few months. However children can continue to improve for up to two years after the illness.



Bring a favourite toy.

The duration of the plateau period (ie the time spent at maximum weakness, before recovery begins) has been found to be the most useful indicator of persisting problems. If the plateau period is longer than 18 days then your child may have some residual problems. An estimated 16% (1 in 6) of children with GBS are left with some residual weakness. Improvement after a 14 days plateau period is usually associated with complete recovery.

GBS is a life-threatening condition and, sadly some children (about 3-5%) may die during the acute phase of their illness. This is usually because of problems with their breathing. However current day medical progress and facilities are becoming so advanced that the mortality rate is reducing all the time.

Recurrence

GBS usually occurs only once. There are isolated cases where recurrence or relapse has occurred. This is rare, especially in children. Doctors have been unable to establish any convincing evidence that GBS may be inherited, so other members of the family will not carry any more risk of the condition than the general public already does.

Variations

As mentioned earlier, not all children conform to the typical pattern. A very small number may follow a more slowly progressive form of weakness which tends to cover months rather than weeks of illness involvement. This is referred to as **chronic inflammatory demyelinating poly[radiculo]neuropathy (CIDP)**; it is very rare in childhood. Another very rare condition in childhood is the Miller-Fisher syndrome. This consists of ataxia (being very unsteady), areflexia (loss of reflexes) and ophthalmoplegia (difficulty moving the eyes) and problems with feeding, swallowing and speaking. It is felt to be a variant of GBS.

Lastly the nerve itself (the axon, rather than simply the nerve sheath) may be involved. This is referred to as acute axonal neuropathy. This is important since recovery takes longer and may not be as complete as for typical GBS.

You may hear of some of these conditions referred to during your child's admission but it remains extremely unlikely any of them would occur.

Who's who?

The team involved in your child's care are:

- you, your child and your family;
- nurses (for the neurology ward and the children's intensive care unit);
- doctors:
 - paediatricians,
 - paediatric neurologists (consultant, senior registrar/registrar, senior house officer),
 - paediatric intensive care doctors,
 - neurophysiologist (who does the nerves conduction study);
- physiotherapists (to help with movement and breathing);
- speech therapist (to help with feeding and communication);
- occupational therapist (to help maximise recovery); and
- dietician.

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JENNY'S STORY



Available from the Support Group is *Jenny's Story*, sensitively written by Dr Jo Wilmshurst and beautifully illustrated by Sean Hatherill.

This book will explain GBS to children suffering from the illness and be helpful reading for other children.

Text by Dr Jo Wilmshurst. First edition June 1998.
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