

# CIDP

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

## **CIDP (Chronic Inflammatory Demyelinating Polyneuropathy)**

A guide for patients, relatives and friends

Second 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](mailto: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

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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.

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LCC Offices, Eastgate**

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# CIDP

## *A guide for patients, relatives and friends*

### Introduction

This booklet has been written for patients who have been told that they may have CIDP (**chronic inflammatory demyelinating poly[radiculo\*]neuropathy**), and for their relatives and friends. It aims to explain accurately and honestly what CIDP is, and hopefully will answer some of the questions you may have. If you do not understand or are worried by any of the information offered here, do ask your doctor to explain.

The degree of severity of CIDP and the way in which it affects people vary enormously from one sufferer to another. There is no typical CIDP. Therefore one general description and one certain prognosis are not possible. This booklet describes symptoms which are common among sufferers.

### What is CIDP?

CIDP is defined thus:

- ‘chronic’ refers to the gradual course of the illness;
- ‘inflammatory’ means there is strong evidence that it is inflammation that causes the nerve damage;
- ‘demyelinating’ means that the damage is primarily to the insulating myelin sheaths around the nerve fibres; and
- ‘poly[radiculo]neuropathy’; ‘poly’ means many, [‘radiculo’ means root,]

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\*‘Radiculo’ is sometimes omitted.

‘neuro’ means nerve and ‘opathy’ means disease; so poly[radiculo]neuropathy means a disease of many peripheral nerves [and their roots (which are the points of origin of the peripheral nerves from the spinal cord)].

CIDP is a very rare disease of the peripheral nervous system involving gradual development of weakness and loss of sensation predominantly in the arms and legs.

The incidence and prevalence of CIDP are very difficult to determine because of its rarity. Various estimates put the incidence at between 75 and 250 people per year in the UK.

The disease may start at any age, but is slightly more common in young adults. It is more common in men than women. For women, relapses are slightly more likely to occur during a pregnancy year. It is not hereditary; ie it is not passed on to children. It is not infectious; ie it is not caught from, or transmitted to, anybody else. It is not a psychiatric or ‘nervous’ disorder.

No-one is sure what causes CIDP. Current research is investigating the role of preceding infections, immunisations and other events before the onset or relapses of CIDP. However, to date there is no general agreement on what causes the disease.

## **Symptoms**

The severity of CIDP is extremely variable and the symptoms experienced vary considerably between patients. Initial symptoms may be vague and confusing to both the patient and the doctor. Subjective symptoms such as fatigue and sensory disturbance are difficult to communicate. In the early stages it may be difficult for the patient to persuade the doctor that there is anything physically wrong.

Early symptoms usually include either tingling (pins and needles) or loss of feeling (numbness) beginning in the toes and fingers, or weakness, so that legs feel heavy and wooden, arms feel limp and hands cannot grip or turn things properly. These symptoms may remain mild and result in only minor disruption the patient’s normal life. Alternatively they may become progressively and gradually worse over a period of several weeks, months or even

years — sometimes, but very rarely, to the extent that the patient is bed bound with profound weakness of the arms.

CIDP usually presents with both weakness and sensory symptoms, sometimes with weakness alone, and rarely with sensory symptoms alone. The arms and legs are usually affected together, the legs more than the arms. Prickling and tingling sensations in the extremities are common and may be painful. Aching pain in the muscles also occurs. Tendon reflexes are usually lost. As the disease becomes more severe, a tremor may develop, usually in the upper limbs. Very rarely patients may develop facial weakness.

## Diagnosis

CIDP can be difficult to diagnose as there is no conclusive diagnostic test for it. The history of symptoms is often vague with varying signs which could be symptoms of a number of conditions. Therefore a long period of time may elapse before a suggestion of CIDP is made.

CIDP is closely related to Guillain-Barré syndrome (GBS), which is also due to inflammation of the peripheral nerves. Symptoms experienced by patients are similar, but GBS is a more acute condition in which symptoms appear rapidly over a period of days or a few weeks. GBS patients usually make a spontaneous recovery over a period of weeks or months.

CIDP is a chronic condition and is only distinguished from GBS by virtue of its pattern of progression. In GBS the low point is reached within four weeks whereas in CIDP the initial progressive phase lasts longer, usually much longer. Some CIDP patients are initially diagnosed as having GBS. Only when the deterioration continues over an extended period, or when one or more relapse(s) occur after a period of improvement, is the illness reclassified as CIDP.

The diagnosis is made primarily on clinical grounds, not laboratory tests. This means that the doctor has to rely on the history and clinical examination fitting into the pattern of CIDP. The doctor will particularly want to know of any recent possible toxin exposure (insecticides, solvents), medication, alcohol intake, tick bites, family history of nerve disease, or symptoms of any coincidental illnesses, such as diabetes (thirst, frequent urination, weight loss)

or arthritis (painful joints). Any of these might lead to a different diagnosis.

Essential criteria for a positive diagnosis of CIDP are:

- progressive weakness in two or more limbs due to a poly[radiculo]neuropathy;
- loss or diminution of tendon reflexes;
- progression for more than eight weeks or recurrence or relapse; and
- evidence of damage to peripheral nerve myelin from nerve conduction tests.

Investigations will include blood tests, usually a lumbar puncture and nerve conduction tests with an electromyogram (EMG) machine, and possibly a Magnetic Resonance Image (MRI) scan. A nerve biopsy may also be performed. In cases where CIDP is associated with an abnormal protein in the blood (Paraproteinaemia) a bone marrow examination and X-rays of the bones may be required.

The lumbar puncture involves lying on one side and having a needle inserted under local anaesthesia between the vertebrae into the sac of cerebrospinal fluid which surrounds the nerve roots. The idea is worse than the procedure really is and it does not usually hurt. The cerebrospinal fluid often 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 EMG is an electrical recording of the muscle activity. 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 of nerve conduction worked out. Usually in CIDP nerve conduction is markedly slowed or even blocked. The test lasts about half an hour. It is only slightly uncomfortable and quite harmless.

The Magnetic Resonance Image (MRI) Scanner is a more recent diagnostic tool and takes X-ray type pictures of the brain and spinal cord (ie of the central nervous system). The procedure involves the patient's upper body being slid into the tunnel-like scanner and remaining absolutely still during the scanning process which lasts about half an hour. It is entirely painless.

MRI scans are used to eliminate the possibility of damage to the central nervous system.

Sometimes a nerve biopsy may also be performed. This involves a small piece of nerve being removed, usually from the side of the heel of the foot, to be examined in the laboratory. This allows the doctor to see any inflammation and the type of nerve damage. Having the biopsy is not painful because local anaesthetic is used, but the skin below may become sore for a week or two afterwards. The patient may be left with some loss of sensation in a very small area on the side of the foot.

## **Progression**

It is helpful to subdivide CIDP into four sub-categories which are characterised by the pattern of progression of the disease. These are:

- ‘subacute’ where symptoms continue to progress and worsen for at least four weeks, but not more than eight weeks before levelling off or improving;
- ‘chronic progressive’ where symptoms continue to progress and worsen for a period exceeding eight weeks;
- ‘chronic relapsing’ where there is more than one episode in which symptoms progress and worsen for a period greater than four weeks; and
- ‘recurrent GBS’ where each bout has a progressive phase of less than four weeks.

Clearly the cutoff points used are somewhat arbitrary.

The most common form of the disease is the chronic relapsing form largely due to the beneficial effects of treatment but sometimes due to spontaneous remissions. About 80% of patients have this form of the disease. About 10% of patients have the subacute disease which plateaus and then disappears spontaneously. Patients with recurrent GBS form only a small percentage of CIDP patients.

Thus some patients only have a single ‘bout’ of CIDP lasting for several months or years, after which a spontaneous recovery may be made. Others

have many bouts in between which spontaneous remission and recovery occurs. After each bout patients may be left with some residual numbness and weakness and sometimes discomfort. For many this will not seriously interfere with their lives, and they are able to continue with or resume their normal occupation. However a very small number are left severely disabled and may be dependent on a wheelchair or even bed bound. There are only a very unfortunate few for whom the disease continues to progress without remission.

## **What is going on?**

The function of the brain is to interpret sensations and initiate movements and other responses. This activity depends on a complex communication system of nerves running to every part of the body via the spinal cord. Each nerve in this communication system can be compared to an electric cable. The inner part of the nerve, the axon, is made of conductive tissue and carries messages or impulses throughout the body — like the wires in an electric cable. The axon is surrounded by a layer of fatty substance, the myelin sheath, like the insulating cover on a cable. The myelin helps the conduction of messages along the nerves as well as insulating and protecting the nerve.

The symptoms of CIDP are due to inflammation and damage to the peripheral nerves and their roots. The peripheral nerves connect the central nervous system to the skin and muscle. CIDP is probably an autoimmune disease, ie one in which the immune system attacks its own body. The most likely mechanism is that the immune cells, called lymphocytes, somehow or other make a mistake and attack the nerves. The main part of the nerve which is attacked is the insulating sheath, or myelin.

The way in which the lymphocytes are tricked into attacking the body is still the subject of research. The lymphocytes may cause the formation of chemicals called antibodies which circulate in the blood and damage the myelin. Attempts to identify these antibodies have so far been only partially successful.

Fortunately the myelin sheath can be replaced within a few weeks or months by the myelin-forming cells, named Schwann cells. If the nerve axons are

damaged these can also regrow, but this is much slower. Research is continuing into the underlying causes and mechanisms of the disease.

## **Treatment**

Treatment of CIDP is usually very effective with about 80% of new cases making a dramatic response to therapy, although there is no one shot curative treatment in the way that antibiotics might cure an infection. Drug treatments are generally thought to work by suppressing the autoimmune response. This in turn reduces the disabling symptoms of the disease. Examples are steroids, immunosuppressive drugs, 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 treatments are available, that demyelinated nerves can repair themselves, and that some patients get better without treatment.

Because of the small number of patients and because most of the treatment methods are quite new, there is limited evidence available of the relative effectiveness of different treatments. Some patients respond to one method of treatment and not to others. There are only a very unfortunate few who cannot be helped by any of these treatments.

## **Steroids**

Controlled trials have demonstrated that steroids are beneficial in CIDP. A wide range of dosage schedules has been used and no work has been addressed to the question of which is best.

The high risks of serious side effects resulting from the prolonged use of high dose steroids are well known. These include osteoporosis (thinning of bones), cataracts, diabetes, hypertension (raised blood pressure), obesity and myopathy (muscle weakness).

If the dosage levels required to control the CIDP appear unacceptably high or unacceptably prolonged, it may be suggested that other immunosuppressive drugs are used.

## **Immunosuppressive drugs**

Clinical experience suggests that immunosuppressive drugs help. These include azathioprine, cyclophosphamide and cyclosporin. Azathioprine is the most widely used in the treatment of CIDP.

The use of these drugs carries the theoretical side effect of increased risk of developing cancer, but in practice this increased risk is very small.

## **Plasma exchange**

Plasma exchange involves the patient being connected to a machine which can separate the blood cells from the fluid or plasma. In an on-line process, blood is continuously taken from the patient, separated, the plasma is discarded, the blood cells are mixed with clean plasma and returned to the patient (the process is not unlike that used in kidney dialysis). At each session about two to three litres of plasma are exchanged. The procedure is usually repeated several times over about two weeks until sufficient plasma has been changed. The procedure is safe and the risks are small. It is not painful. However some patients find that it leaves them feeling tired for a day or two.

Clinical trials have demonstrated the benefit of plasma exchange for CIDP. For some patients it allows control of the disease to be maintained when immunosuppressive drugs are insufficiently effective. Some patients however do not appear to respond to plasma exchange.

## **Immunoglobulin**

There is increasing evidence of the effectiveness against CIDP of intravenous infusions of immunoglobulin (also called gamma globulin or antibodies). Antibodies usually react with and neutralise germs which get into the

body. These are 'good' antibodies. Sometimes antibodies attack the body itself and these 'bad' antibodies, or autoantibodies, may cause CIDP. However there are also anti-autoantibodies, which block these bad antibodies. It may be these anti-autoantibodies in immunoglobulin which help.

Whatever the explanation, some people with CIDP do seem to get better after having immunoglobulin. Research is going on to find out which patients.

It is given by infusion into a vein, usually every day for five days. Each infusion takes about five hours. The immunoglobulin used in the UK has an excellent safety record. Abroad there have been very rare cases of transmission of hepatitis but considerable care is now taken in the purification and removal of any viral particles which reduces this risk to the absolute minimum. With any blood product there is always a slight risk of transmission of a new infection such as Creutzfeldt Jakob disease (CJD) which has received a great deal of recent publicity. For this reason immunoglobulin from British Donors is not currently being used as a source for the manufacture of immunoglobulin until there is confirmation that there is no risk associated with it. This extra safeguard should reduce the concerns of anyone receiving this very effective treatment. There is a rare (about 1 in 40,000) risk of serious allergic reaction at the start of each infusion, so careful monitoring is essential. Some patients only need one course. Others need repeated courses.

## **Physiotherapy**

Physiotherapy has an important role to play in the assessment and management of CIDP. It helps to maximise a patient's physical potential, particularly where weakness is the predominant problem.

The aims of physiotherapy are to:

- maximise muscle strength and minimise muscle wastage by exercise using strengthening techniques;
- minimise the development of contractures (or stiffness) around joints; a physiotherapist can advise on passive stretching techniques to help maintain full range movement at joints;

- facilitate mobility and function; sometimes, if muscles are very weak, function can be improved by the use of splints and
- provide a physical assessment which may help in planning future management.

# **Living with CIDP**

## **Coping with uncertainty**

CIDP may follow a pattern of relapses and remissions or a more gradual increase in symptoms. During a relapse new symptoms occur or old symptoms which had previously subsided may recur. Relapses can last for several months and may be relatively slight or quite severe. A remission occurs when the symptoms experienced during the relapse disappear either partially or completely over a period of time which may last weeks, months or even years.

CIDP does not always have these patterns of being ‘better’ or ‘worse’; sometimes symptoms can gradually increase over a period of many years and it may be difficult to identify ‘better’ or ‘worse’ times.

It is impossible to predict with certainty how CIDP is going to affect an individual in the future. The pattern of relapses and remissions varies greatly from person to person. A period of relapse can be very disturbing but many people make a good recovery. Coping with this uncertainty is one of the most difficult aspects of ‘living with CIDP’. You should try and accept this variability without getting too worried about it.

## **You and your family and friends**

A diagnosis such as CIDP of a chronic condition with an uncertain prognosis, may well throw a strain on family and other relationships. You may find

it difficult to accept help when you need it, or your family and friends may feel that they cannot give help or become overprotective toward you. It is difficult to carry on family life as if nothing has happened. Everyone concerned may have to take on new roles. If you and your family and friends are able to speak openly and honestly with each other you will probably find that you are able to help each other through difficult times with the result that the bonds are strengthened.

Instinctively children are aware that something is wrong and that you are worried. It is important that their questions are answered as and when they occur. Older children can become surprisingly mature and a source of strength. Trying to keep your problems to yourself will not spare them any anxiety.

## **You and your doctor**

It is important to build a good relationship with your doctors, both GP and specialist. Because of the rarity of the illness, many doctors will not have encountered it before. The symptoms are difficult to describe and may not be taken seriously at first. Each case of CIDP is different, and relapses, if they occur, may bring new symptoms and problems. Because of the variability in severity and progression of the disease, the doctor will not be able to give you a definite prognosis.

Although there is not one single overall treatment for CIDP, there is much that your doctor can do to help. Each person responds in different ways to different treatments. A period of experimentation with different treatment regimes is likely to be necessary in order to discover the regime which is most appropriate for you.

## **Attitude to life**

It is important to be as positive as possible about everything. Our emotional state plays a large part in our health and although the norms of life may have to change for a while, the majority of patients with CIDP can expect a good quality of life.

Modification of one's lifestyle may be necessary but it is better to emphasise strengths, undertaking what can be achieved rather than failing to achieve the impossible. It is a natural reaction to become frustrated but the acceptance and understanding of the problem is more than half the battle. Addressing the problems of CIDP can be seen as bringing a new challenge.

Being positive can take a lot of effort, determination and even courage and can be helped by a similar attitude in those that support and help you.

## **What you can do to help yourself**

You should follow as healthy a lifestyle as possible. This will help to prevent other illnesses and infections which have been shown to trigger relapses.

A nutritionally balanced diet will ensure you are getting all the vitamins and minerals you require. There is no evidence of any special dietary requirements for CIDP sufferers. It is sensible to keep your weight down, since more weight is more difficult for weak legs to carry.

Regular exercise is important for overall health and should be taken according to individual limits and capabilities. Over exertion causes fatigue. However a little regular exercise will help to minimise muscle wastage and give you a good feeling of wellbeing. Any form of exercise that you enjoy and can comfortably follow will prove beneficial. Ask your physiotherapist to show you.

Adequate rest periods are essential to avoid fatigue. Stress and tension may irritate the symptoms of CIDP and therefore relaxation will allow you to unwind and 'recharge'.

Some patients find it useful to record their progress in a diary so that they can discuss changes of treatment in the light of their recent progress. Others find that this can increase their anxiety about the disease and is counter-productive.

Original text by Eileen Evers and Professor Richard Hughes.

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