

# GUIDE

## *Chronic Inflammatory Demyelinating Polyradiculoneuropathy*

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

### **Introduction**

This guide 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 or specialist to explain.

### **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,] 'neuro' means nerve

\*'Radiculo' is sometimes omitted.

and 'opathy' means disease; so poly[radiculo]neuro-pathy 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 rare disease of the peripheral nervous system which affects about 500 people in the UK at any one time. It has a number of different forms which vary enormously in severity. Because of its rarity it is often difficult to diagnose and it may take some time to be referred to a specialist who would recognise the pattern of disease and start appropriate treatment. It is not an inherited disease and it is very rare for more than one member of a family to get the condition. It is possible that the tendency to react abnormally to infection by producing antibodies that harm nerves as well as fighting infection may be more common in some families but there are so many other factors that are needed to start off the disease that in practice it is not inherited. There is no evidence that you can catch CIDP from anyone else and it is not infectious in that sense. Nor is it a 'nervous' or psychiatric disease. It can start at any age and is slightly more common in men than women.

No-one is sure what causes CIDP. Quite a few patients are aware of an initial infection that triggered the disease. It is possible that vaccinations may trigger the disease although this does seem to be a very low risk with current vaccines. (The GBS Support group has advice on the safety of vaccinations in CIDP which tries to clear up this very confusing area.)

Over the last few years the understanding of CIDP has increased and it is now believed that CIDP is the name given to a chronic disease of the peripheral nerves that reflects a number of different ways in which nerves can be damaged. Probably the most common is a disorder in which the immune system attacks the outer insulating nerve layer leading to weakness and altered sensation that comes on rather slowly often over a few months. Weakness is usually the most prominent complaint and can be so severe as to confine patients to a chair or bed.

Variants of the disease can occur in which the weakness pursues a relapsing course rather than being slowly progressive and some patients develop only sensory symptoms. These variants are named according to the pattern of the disease course as chronic relapsing CIDP and sensory CIDP. There is also a sub-acute form of the disease known as sub-acute CIDP.

There are other forms of CIDP in which the axons or central conducting fibres are mostly damaged (axonal CIDP) and there are patients in whom the CIDP is associated with abnormal proteins in the blood or paraproteinaemias (CIDP with paraproteinaemia). A very closely allied disorder which is probably a variant of CIDP is called multifocal motor neuropathy and is characterised by block of electrical conduction down the motor peripheral nerves. This disorder can sometimes be mistaken for motor neurone disease but usually responds very well to immunoglobulin treatment. Another variant of CIDP is called multifocal acquired demyelinating sensory and motor neuropath (MADSAM). This disorder is usually asymmetrical and worse in the arms but is very similar to CIDP in symptoms and response to treatment.

Since we do not completely understand the cause of the disease it is possible that some of these forms might be different stages of the same process but it is likely that the mechanisms of nerve damage are different in the different forms of the disease.

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

## **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. Arms and legs are usually affected together. These symptoms may remain mild and result in only minor disruption of the patient's normal life. Alternatively they may become progressive 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. Occasionally patients have only sensory symptoms. 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. Tremor of the hands can sometimes occur. Sometimes patients may develop weakness of the voice or face, but this is usually mild compared with the symptoms in the arms and legs. Breathing and swallowing are only very rarely affected.

## **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.

A diagnosis of CIDP requires the following:

- Weakness of at least two limbs;
- Complete or partial loss of tendon reflexes;
- Progression over eight weeks or relapses;
- Evidence of myelin damage in the peripheral nerves from nerve conduction studies.

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 patients with CIDP develop weakness acutely in much the same way as patients with GBS but instead of stabilising and then improving they go on to get worse for several months. This slowly progressive course points to the real diagnosis of CIDP.

A diagnosis of CIDP is usually made on clinical grounds but with evidence from nerve conduction studies of demyelinating neuropathy. Examination of the cerebrospinal fluid (lumbar puncture) is usually required to demonstrate a raised CSF protein. It is important to rule out other diseases that can cause demyelinating neuropathy by a thorough examination and a detailed history. Examination of other family members may be necessary to completely rule out an inherited neuropathy. The history will pay particular attention to possible toxins or drugs that could cause neuropathy of which amiodarone is probably the most likely to cause diagnostic difficulty. Other diseases such as diabetes, arthritis or hepatitis will be important as will alcohol intake.

## **Nerve conduction studies**

This test, which is sometimes called EMG (electromyogram), consists of stimulating the peripheral nerves with a small electrical current in order to assess the speed with which electric impulses can pass down the nerve. This usually results in a sharp jolt of the muscle concerned.

A reduction in this speed can suggest damage to the myelin insulating material that ensures nerves conduct electricity quickly. Some people find this test uncomfortable but it is quite harmless and cannot damage the nerves.

## **Lumbar puncture**

A lumbar puncture consists of lying on one side bent double as much as possible in order to open up the spaces between the vertebrae. A doctor then inserts a very fine needle under local anaesthetic into the sac that holds the spinal fluid. This procedure is very similar to an epidural but has fewer complications, and does not usually hurt.

## **Magnetic resonance scan**

MRI scans are now widespread in medicine and consist of using a very strong magnet to take a picture of the nerves and brain. In CIDP an MRI scan is frequently employed to rule out compression of nerve roots by slipped discs. As with all MRI scans the procedure is harmless but does involve lying in a tunnel within the scanner which some people dislike.

## **Nerve biopsy**

Sometimes a nerve biopsy is needed to be certain of the diagnosis. This is performed under a local anaesthetic which is injected into the foot behind the ankle. A small sensory nerve is removed for examination under the microscope. The procedure does not hurt but there is usually a small numb patch on the side of the foot once the nerve has been removed.

## **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 auto immune disease, ie one in which the immune system attacks its own body. In CIDP both sensory and motor nerves are usually involved but in rarer types of the disease either just motor or just sensory nerves are involved. Where both motor and sensory nerves are involved the damage seems to occur because of scattered areas of inflammation within the nerves. This is thought to occur because of stimulation of the immune response to recognise and damage normal nerve components. This stimulation seems to occur following certain infections which fool the lymphocytes in the blood into treating normal nerve as if it was a foreign germ and trying to destroy it. The body tries to repair the damage with varying degrees of success. Drug treatment in CIDP is designed to repair the immune response and in some cases suppress the over enthusiastic immune process.

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 having 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, 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.

There is no doubt that most patients will improve with steroids but unfortunately if high doses are required many patients will experience some side effects. Many of these are minor but patients can develop osteoporosis (thinning of the bones), cataracts, diabetes, hypertension (raised blood pressure) weight gain and muscle weakness.

Many neurologists like to combine steroid therapy with a “steroid sparing drug” designed to suppress the immune system and allow a lower dose of steroids to be used. A number of these drugs are available and include azathioprine, methotrexate, cyclosporin, mycophenolate and cyclophosphamide. These drugs all collectively can suppress the white cell count and therefore have to be carefully monitored by regular blood tests and some require tests of breathing or kidney function. The use of these drugs carries the theoretical side effect of increased risk of developing cancer, but in practice this increase 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. This process is not painful but can be tiring and may take several hours. Plasma exchange is usually performed two to three times a week for two weeks. The effect of the treatment usually only lasts for a few weeks and therefore it needs to be combined with something else or repeated regularly. There are some problems with getting the lines into patients with poor veins and bleeding or infection can occur at the sites of the line insertion.

## **Intravenous immunoglobulin**

Intravenous immunoglobulin has become a common treatment for CIDP and its effectiveness is supported by clinical trials. It has been used in many thousands of patients throughout the world for at least a decade. The immunoglobulin contains many thousands of antibodies derived from healthy donors and the exact way it works is not known. One way it may work is to prevent damaging antibodies in the blood stream from binding to their target but there are almost certainly a number of other beneficial functions of the immunoglobulin.

Because it is derived from a blood product, immunoglobulin can cause allergic reactions such as skin rash or fever. Serious reactions are very rare but the treatment is usually monitored in hospital. It is given by a slow infusion into a vein in the arm and a typical course of treatment would involve five days of infusion lasting a few hours each day. Some patients are able to tolerate a higher dose or more rapid infusion and can have treatment given on one or two days.

With any blood product there is a slight risk of transmitting a new infection such as Creutzfeldt Jakob disease (CJD). For this reason blood for immunoglobulin is not obtained from British donors but from areas of the world where Mad Cow Disease (BSE) is very rare. No conclusive evidence exists that immunoglobulin could transmit CJD but every precaution is being taken to reduce any possible risk to an absolute minimum.

## **Physiotherapy and occupational therapy**

Physiotherapy and occupational therapy both have 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 supports;
- provide a physical assessment of muscle strength, which plays an important part in assessing response to treatment and in planning future management.

## **Living with CIDP**

### **Anxiety**

In the early stages of CIDP, especially before diagnosis, when symptoms are getting progressively worse, many people feel extremely frightened. What have I got? How bad is this going to get? These feelings are quite normal and should be understood by all those around you — family, friends and medical staff. Usually this anxiety diminishes after diagnosis when you understand more about the condition, treatment options have been explained and treatment may have been initiated. Some people find it helpful to be in touch with other sufferers. The GBS Support Group runs a CIDP network. Members of the Network, who are all CIDP sufferers or carers, exchange contact details and a brief outline of their CIDP experiences. The idea is that anyone in the Network can contact anyone else for mutual support and to exchange experiences and information. Information can also be obtained from the Group's Web site [www.gbs.org.uk](http://www.gbs.org.uk) which has an active discussion forum where CIDP sufferers support one another.

### **Attitude to life**

Depression is also a problem for many CIDP sufferers. Those who have previously led very active and busy lives find it very difficult to adapt to new circumstances. Some people feel that life has been unfair (Why me?). Some people lose hope that things will ever be any better. These problems can seem overwhelming especially when you are physically weak and feeling fatigued. All these feelings are understandable. It usually helps to talk about them.

It is important to be as positive as possible about everything. Our emotional state plays a large part in our health and although 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.

## **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 and frightening 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.

## **For carers and friends**

For carers looking after someone with CIDP, life can be very difficult. Carers must balance the needs of the sufferer with those of other family members, whilst at the same time keeping 'normal' life going for themselves — maintaining a job and looking after the family home. Many carers feel frustrated at not being able to 'do' more to make a loved one better. It can feel as if CIDP has taken over the whole family. Most carers find it important sometimes to take time out for themselves. A great deal of patience and understanding is required from everyone.

For friends of a CIDP sufferer there are a lot of practical things you can do to help — help with housework, shopping, etc, help with transport to and from hospital and doctor's appointments, help with children, taking them to school, etc, — the list is endless. It is also important to 'be there' to allow your friend to talk about their fears and frustrations. However the most important thing is to remember that, although what they can do may have changed, at least temporarily, inwardly a CIDP sufferer is still the same person as before.

## **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. When a relapse occurs this can be a period of great anxiety. It helps enormously to know that your doctors are familiar with your condition and know what is the most appropriate treatment for 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 well-being. 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.

Fourth edition April 2004. Revised by Dr John Winer and Eileen Evers.

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.

*giftaid it*

GBS Support Group, LCC Offices, Eastgate, Sleaford, Lincs, NG34 7EB

Tel: 01529 304615 E-mail: [admin@pbs.org.uk](mailto:admin@pbs.org.uk) Web site: [www.pbs.org.uk](http://www.pbs.org.uk)

© GBS Support Group

