



# CIDP & the associated chronic variants

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



## **Chronic Inflammatory Demyelinating**

**Poly(radiculo)neuropathy (CIDP)** is a rare autoimmune condition of the peripheral nervous system. As many as 650 people are diagnosed with CIDP each year in the UK. Studies have shown that CIDP:

- has several different forms which vary in severity
- is not hereditary or infectious
- is not a psychiatric condition
- can start at any age and is slightly more common in men than women

CIDP is closely related to the acute condition, Guillain-Barré syndrome (GBS, also known as AIDP), and is only distinguished from GBS by its pattern of progression. In GBS the low point is reached within four to six weeks whereas in CIDP the initial progressive phase usually lasts 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.

## **Paraproteinaemic Demyelinating Neuropathy (PDN) is**

sometimes described as:

- chronic demyelinating neuropathy associated with a benign paraprotein
- CIDP associated with paraprotein
- CIDP with paraproteinaemia

Antibody-producing bone marrow cells go out of control and produce large numbers of the same antibody. The antibody (or immunoglobulin) sometimes damages nerve fibres causing a

peripheral neuropathy. Some doctors regard the clinical, electrophysiological and pathological features of the demyelinating paraproteinaemic neuropathies and of CIDP as closely similar and almost indistinguishable.

These neuropathies are usually late-onset in terms of age and are mixed motor and sensory, although the severity of sensory loss tends to be greater compared with CIDP. There is usually more pain but less severe weakness and impairment. Most patients respond to corticosteroids, cytotoxic drugs, or plasma exchange.

**Multifocal Motor Neuropathy (MMN) or MMN with Conduction Block (MMNCB)** is sometimes thought of as a rare variant of CIDP. However, there are differences that are more prominent than the similarities. MMN patients commonly have asymmetric weakness of the distal (far) muscles, while in CIDP, proximal (near) symmetric weakness is more common. The remitting and relapsing course that may occur in CIDP is uncommon in MMN. Patients with MMN rarely have significant sensory symptoms, unlike CIDP. Increased protein level in the cerebrospinal fluid of MMN patients is rare. Treatment with IVIg or cyclophosphamide is usually effective.

**Lewis-Sumner syndrome** is also known as **MADSAM** — Multifocal Acquired Demyelinating Sensory and Motor neuropathy. It is a chronic condition with similarities to Multifocal Motor Neuropathy but with enough differences, especially in treatment, to have acquired its own definition. Some report it to be an asymmetrical variant of CIDP. MMN and MADSAM respond to IVIg. Some MADSAM sufferers respond to prednisolone whilst most MMN sufferers do not.

**Chronic axonal neuropathies** are common, particularly as a result of diabetes or alcoholism. However, the medical literature does report cases of immune-mediated chronic axonal neuropathy though there are suggestions that this is a secondary result of myelin damage that ultimately appears to be the primary cause of the condition.

### **Sub-acute Inflammatory Demyelinating**

**Poly(radiculo)neuropathy (SIDP)** - GBS is defined when the nadir (worst point) occurs within four weeks of first symptoms. Usually it is much less. CIDP is defined when the nadir comes after eight weeks. Usually it takes much longer. An illness peaking after four weeks but before eight weeks may be called subacute and will be treated as CIDP or GBS depending on which it best resembles.

## **The diagnosis**

### **What causes CIDP?**

No-one is sure what causes CIDP. Quite a few patients are aware of an initial infection that triggered the condition. It is possible that vaccinations may act as a trigger although this does seem to be a very low risk with current vaccines.

### **What are the symptoms?**

The symptoms experienced vary considerably between patients and may be vague and confusing to both the patient and the doctor. Subjective symptoms such as fatigue and sensory disturbance are difficult to communicate.

Early symptoms usually include

- tingling (pins and needles)
- loss of feeling (numbness) beginning in the toes & fingers
- weakness, affecting arms and legs (usually 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.

### **How is CIDP diagnosed?**

CIDP can be difficult to diagnose as there is no single, conclusive diagnostic test for it. The symptoms are often vague and can be produced by a number of different conditions. Therefore, a long period of time may elapse before a diagnosis 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 or relapse eight weeks or more after initial disease onset
- evidence of myelin damage in the peripheral nerves from nerve conduction studies

A diagnosis of CIDP is usually made on clinical grounds but with evidence from:

- nerve conduction studies
- lumbar puncture
- MRI scan
- nerve biopsy
- ruling out other diseases that can cause demyelinating

neuropathy

- family history to completely rule out an inherited neuropathy
- contact with possible toxins or drugs that could cause neuropathy
- other conditions - diabetes, alcohol dependency, arthritis or hepatitis

### **A bit more about the tests**

The **Electromyogram** is an electrical recording of muscle activity and is a very important part of making the diagnosis. The test usually last about half an hour and some patients find the electrical stimulation rather uncomfortable but it is entirely harmless. The **lumbar puncture** tests for protein levels in the spinal fluid and involves lying on one side and having a needle inserted into the base of the spine under local anesthetic. The **MRI** scan is used to rule out compression of nerve roots by slipped discs. A **nerve biopsy** is when a small sensory nerve is removed for examination under local anesthetic.

## **Treatment and management**

### **Is there a treatment?**

Treatment of CIDP is usually very effective, with about 80% of new cases having a dramatic response to therapy. Although some patients go into a long-term remission after a short course of treatment, many require long term treatment of one form or another. Drug treatments are generally thought to work by suppressing the autoimmune response, which in turn reduces the disabling symptoms of the disease.

Examples of treatments are:

- steroids
- immunosuppressive drugs
- plasma exchange
- intravenous immunoglobulin
- subcutaneous immunoglobulin

Some patients respond to one method of treatment and not to others. However, there are a few who cannot be helped by any of these treatments. Suppressing the immune response cannot be undertaken lightly because it can increase the risk of infections. The decision whether to try these treatments must be tailored by the doctor to the individual needs of each patient. However, it may be reassuring to know that treatments are available, that demyelinated nerves can repair themselves, and that some patients get better without treatment.

## **Information about the treatment and management**

### **Steroids**

Controlled trials have demonstrated that steroids are beneficial in CIDP. A wide range of dosage schedules has been used but it is not clear 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 (weakening of the bones), cataracts, diabetes, hypertension (raised blood pressure) weight gain and muscle weakness.

### **Plasma Exchange**

Plasma exchange involves the patient being connected to a machine which can separate the blood cells from the fluid or plasma. 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.

## **Immunoglobulin**

Intravenous immunoglobulin (IVIg) has become a common treatment for CIDP and its effectiveness is supported by clinical trials. It has been used to treat many thousands of patients throughout the world for at least a decade. The infusion contains many thousands of antibodies derived from healthy donors and the exact way it works is not known.

### **How is immunoglobulin given?**

Usually IVIg is given in hospital, usually in a day care unit. It is given through a drip intravenously and the rate, dose and time are calculated individually for each patient. If the treatment is successful it may be given on repeated occasions, often every 4-8 weeks. In long term use in some parts of the UK it is possible to receive IVIg at home given slowly under the skin (see **Subcutaneous Immunoglobulin** below). Typically, this is more suitable for patients who are well established on immunoglobulins and whose total monthly dose is not too high.

### **Are there any side effects of IVIg?**

As with all treatments, side effects can occur with IVIg, although usually these are minimal and do not require the treatment to be stopped.

Transient side effects, which often respond to changes in the rate of administration of the infusion, include headache and low blood pressure. It is helpful if you drink plenty of fluid whilst you are receiving the IVIg. More rarely, a rash can develop.

IVIg thickens the blood slightly so particular consideration of its use is given to patients with kidney failure, previous heart disease, strokes or blood clots. Very rarely such severe complications can result from IVIg use.

### **Subcutaneous Immunoglobulin**

Subcutaneous immunoglobulin (SCIg) has been developed more recently than IVIG. With SCIg, immunoglobulin is delivered by a needle into the fatty tissues under the skin, where it enters the circulation slowly over a few days. There isn't much room under the skin, so the dose of immunoglobulin given is smaller than with IVIg. For this reason, SCIg is usually given every week. Nearly everybody on SCIg learns how to have treatment at home, with each session lasting up to about two hours.

### **Physiotherapy and Occupational Therapy**

Physiotherapy and occupational therapy both have an important role to play in the assessment and management of CIDP. They help 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

## **What happens next**

### **Going home**

Not everyone with CIDP will require a stay in hospital. However, if you do need to be admitted, then leaving hospital or a rehabilitation centre and heading home can be daunting and take a while to arrange. You may need equipment to help with everyday tasks, your home may need adaptations or you may need a care package in place to help. There are many people and organisations that can help with this starting with the occupational therapists and your care team. Your family can also be a great help in getting the information together and speaking to organisations that have in depth knowledge of what help is available.

### **Assessment & care plan**

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

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

The type of support that might be in a care plan

- community care services to allow you to live in your home

- NHS continuing healthcare
- NHS funded nursing care
- rehabilitation
- equipment provision
- support from voluntary agencies

## **Benefits**

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

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

### **Personal Independence Payment (PIP) is:**

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

### **Attendance Allowance (AA) is:**

- for people aged 65 and over who have a health condition which has lasted at least six months

- not affected by any income or savings you have, is tax-free and payable alongside other benefits (except Disability Living Allowance or Personal Independence Payments). You do not need to have paid national insurance contributions
- Entitlement is based on care needs resulting from how your health affects your everyday life

### **Disability Living Allowance (DLA) is:**

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

### **Disability Living Allowance for Children is:**

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

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

- is payable to people unable to work because of ill health or disability.
- requires a medical certificate ('fit note') from your GP to make a claim.

- You will be required to fill out a medical questionnaire, attend a medical assessment and a work-focused interview. This is intended to determine your capability to work. It is possible to challenge the decision.

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

### **General prognosis**

Treatment of CIDP is usually very effective with about 80% of new cases having a dramatic response to therapy. Although some patients go into a long-term remission after a short course of treatment, many require long term treatment of one form or another. Drug treatments are generally thought to work by suppressing the autoimmune response, which in turn reduces the disabling symptoms of the disease.

### **Hygiene and cleanliness**

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

### **Teeth**

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

can help. Using an electric tooth brush can be helpful if you have residual weakness in your hands.

### Other sources of support

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

### **Diet**

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

### **Exercise**

Some patients may require physiotherapy, occupational therapy (OT) and speech and language therapy. These can play a vital role in the rehabilitation process as well as maximizing functional ability. At some point during rehabilitation the rate of recovery may plateau and it is often at this point that patients will be discharged from all the support services on which they may have relied. It is also possible that patients may be placed 'on review'. This means that you may be followed up at regular intervals and can telephone for advice in-between but don't attend the clinic as often as you did before.

Exercise can help to improve your muscle strength and reduce your overall sense of fatigue. There are also general benefits of exercise in boosting the immune system, helping your heart and

lungs remain healthy and making you feel better about yourself. However, it may take weeks or even months before you feel the benefit of exercise so it is important to pace yourself. Therefore, you should be encouraged to seek advice on whether and how to start regular exercise.

## **Pain**

Pain may never be a problem but can occur and some patients experience painful pins and needles or other unpleasant sensations, such as burning feelings in the hands and feet. The problem does tend to resolve as recovery proceeds.

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

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

### Other sources of support

Pain Concern	<a href="http://www.painconcern.org.uk">www.painconcern.org.uk</a>
Pain Society	<a href="http://www.britishpainsociety.org">www.britishpainsociety.org</a>
Welsh Pain Society	<a href="http://www.welshpainsociety.org.uk">www.welshpainsociety.org.uk</a>
Pain Relief Foundation	<a href="http://www.painrelieffoundation.org.uk">www.painrelieffoundation.org.uk</a>

## **Wellbeing**

### **Sexual relationships**

GBS, CIDP and associated inflammatory neuropathies can bring on problems in any relationship, sexual relationships are not excluded. Dealing with a long term illness or disability can put a great strain on a relationship, particularly when one partner is partially or totally dependent on the other. Even without the actual



physical disability, the emotional upheaval can interfere with a couple's sex life and this can be difficult to talk about. This can mean that the once close, intimate relationship can become distant and stressful for both partners. Help is available so speak to your GP or a relationship counsellor.

### Other sources of support

Relate [www.relate.org.uk](http://www.relate.org.uk)  
Sexual Dysfunction Association [www.impotence.org.uk](http://www.impotence.org.uk)

### **Emotional issues**

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

There is help available and you can speak to your GP or neurologist about this. They will be able to arrange suitable help for you.

### Other sources of support

Wellbeing [www.nhs.uk/Conditions/stress-anxiety-depression/.../improve-mental-wellbeing.aspx](http://www.nhs.uk/Conditions/stress-anxiety-depression/.../improve-mental-wellbeing.aspx)  
Mindfulness [www.bemindful.co.uk](http://www.bemindful.co.uk)  
NHS [www.nhs.uk/Conditions/stress-anxiety-depression](http://www.nhs.uk/Conditions/stress-anxiety-depression)  
MIND [www.mind.org.uk](http://www.mind.org.uk)

### **Immunisation**

Little is known about the risks of immunisation in CIDP. However, it is impossible to deny that relapses sometimes happen after immunisations in CIDP. In many other neurological diseases, for instance multiple sclerosis, there is more information and influenza vaccine is considered safe.

### **What should I do?**

This always depends on your individual circumstances. You must balance the benefits of the vaccine against the unknown but probably small risk of the vaccine causing a relapse.

**You should always discuss this with your own doctor.** Ask your doctor if the vaccine is necessary. The following are common questions:

### **I am NOT on steroids, plasma exchange, azathioprine or other immunosuppressive drugs: should I be immunised?**

Some immunisations are more important than others. For example, most people have already been immunised against tetanus, and boosters may not be essential. However, if you have not been immunised within the past 5 years and cut yourself so that dirt gets into the wound then the balance of risks may change in favour of receiving the vaccine.

### **I AM on steroids, azathioprine or other immunosuppressive drugs: should I be immunised?**

Theoretically your risk of developing infections like a serious case of influenza is greater because you are on these drugs. However, your risk of having a relapse of CIDP is also probably less because you are on them. The balance of evidence may therefore be more in favour of having, for instance, an annual influenza vaccine.

## **I am on intravenous immunoglobulin: should I be immunised?**

Intravenous immunoglobulin probably makes you less likely to have infections so the need for immunisation is less. Also intravenous immunoglobulin probably makes immunisations less effective. If you decide to be immunised, theoretically it is probably better to do this half way between your intravenous immunoglobulin courses. Reminder: The decision to have a vaccine depends on your individual circumstances and you should always discuss this with your own doctor. You might wish to show him or her a copy of this guideline.

## **Advice for the carer**

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

- Gather support from family and friends. Invite help from the local social services/social work department both practical and financial
- Contact a local caring organisation providing support services in your area. They will often help to bath and dress patient, providing a respite so that the carer can go shopping or have a bit of time of their own.
- Take a rest from your duties and allow yourself some personal space. Go for a walk, listen to relaxing music, visit friends etc. Generally, take care of yourself, eat healthily, and get plenty of sleep. When friends or relatives visit the patient, take this as an opportunity to have a break and use this time to do something for you.
- Take the pressure off by putting some activities on hold.

- Be mindful of the patient's limitations.
- Talking is therapy and you may also find it useful to speak to an external source: friends, relatives, caring organisations, **GAIN** etc.
- Get organized. Investigate benefit entitlements with the hospital social worker and/or Social Services/Social Work Department. Liaise with the hospital occupational therapist (OT) and physiotherapist about equipment arrangements.
- Be temperature conscious if the patient is suffering from lack of sensation, ie run and test the bath water. This also applies to the cooker, iron etc. as there may be no sensation and a patient can get burnt or scalded very easily.
- Taste buds may be affected for a while, so prepare meals to suit the patient. Vitamin supplements can be included if a balanced diet cannot be achieved.
- Beware of falls brought about by weakness or unsteadiness.
- Be mindful of potential accidents resulting from weakness and/or numbness (ie dropping things). Care should be taken when the patient is using hot appliances, such as when cooking or ironing. A microwave oven is a very convenient, safe way of preparing food.
- Help the patient with daily exercises. Ensure that everything is done in moderation and that the patient does not start rushing around too soon. Encourage the patient to talk openly about his/her experiences and fears.

Other sources of support:

Carers UK

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

Carers Scotland

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

Carers Northern Ireland

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

Carers Wales

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

The Carers Association ROI

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

The Princess Royal Trust for Carers

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

Crossroads Association

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

Age UK

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

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

### Acute

**GBS** Guillain-Barré syndrome

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

**MFS** Miller Fisher syndrome

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

**AMSAN** Acute Motor Sensory Axonal Neuropathy

### Sub-acute

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

### Chronic

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

**CIAN** Chronic Idiopathic Axonal Neuropathy

**CMFS** Chronic Miller Fisher syndrome

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

**MMN** Multifocal Motor Neuropathy

**MMNCB** Multifocal Motor Neuropathy with conduction block

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

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

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

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



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

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

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