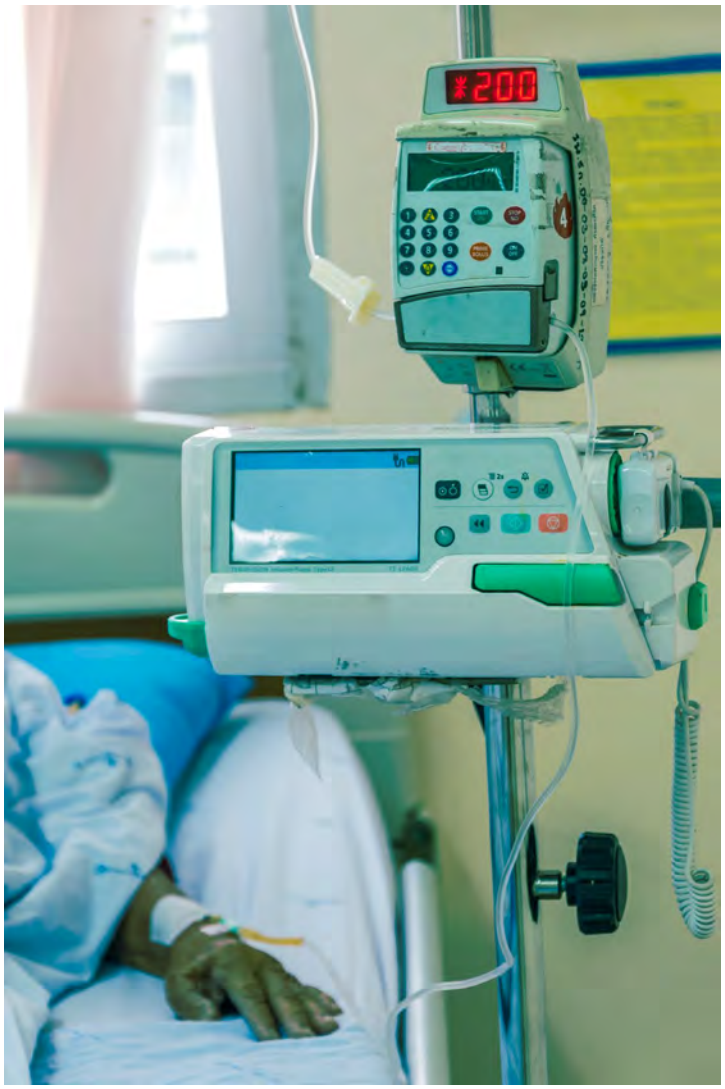
Dedicated to helping people affected by Guillain-Barré syndrome, CIDP and the associated inflammatory neuropathies



CIDP and related conditions

About this booklet

Most people have never heard of CIDP (chronic inflammatory demyelinating polyneuropathy) until they find themselves affected by it. The symptoms can vary significantly, and may be mistaken for those of other neurological conditions, making diagnosis of this rare autoimmune disorder potentially difficult. The road to recovery is often long and the future uncertain. This can be a confusing and frightening time, but GAIN is here to provide information and support for as long as it is needed.

In addition to our information booklets and factsheets, we can provide a hospital communications book for people who are ventilated and non-verbal for a time, help with the cost of frequent hospital visits for a family member, and arrange contact with a peer support volunteer who has had first-hand experience of living through this distressing condition. We also have a website with information to read and download, and a friendly Facebook community you can join.

This guide has been approved by the GAIN Medical Advisory Board and provides an accurate account of the conditions. Not all content will apply to you as symptoms, severity and the extent and timescale for recovery will vary from one person to another. If you need more specific information about your condition, please talk to your consultant or GP. If you need further help from GAIN, please get in touch or visit our website.

Guillain-Barré & Associated Inflammatory Neuropathies (GAIN)
Glennys Sanders House
Pride Parkway
Sleaford
Lincolnshire

office@gaincharity.org.uk

Monday to Friday 9am – 3pm

www.gaincharity.org.uk

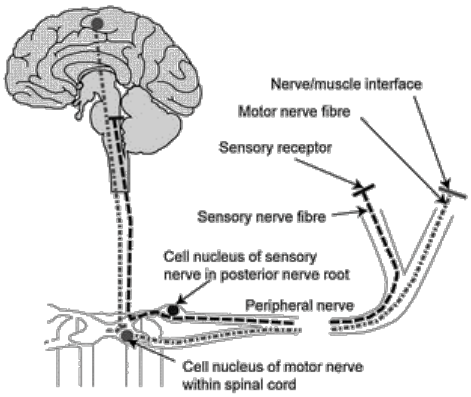
Registered charity 1154843 & SCO39900

Contents

Introduction

Before we explain the chronic conditions in more detail, it is worth understanding a little about how the peripheral nerves work and how that affects you.

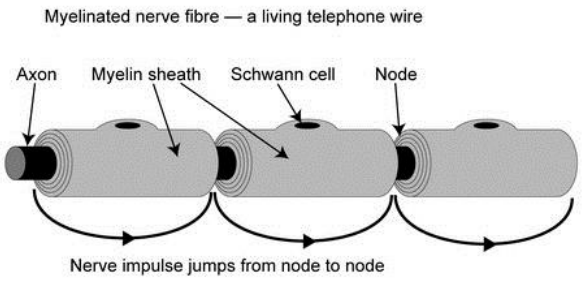
The peripheral nerves connect the central nervous system (the brain and the spinal cord) to the periphery (the sensory receptors and muscles). An illness of the peripheral nerves is called a 'peripheral neuropathy'.



Peripheral nerves are made of bundles of nerve fibres, which can be regarded as living telephone wires. They are kept alive by their cell bodies. The cell bodies of the motor nerves lie in the spinal cord within the spinal column or around the base of the brain. The cell bodies of the sensory nerves are in bunches, called ganglia, connected to the nerve roots on the back of the spinal cord or brain stem.

The motor nerve cell body has a long fibre called an axon, which extends from the central nervous system to the muscles. The longest axons can be as much as a metre in length, for example the nerves to the muscles in the feet. The connection between the motor axon and the muscle fibre is a specialised nerve ending, which contains tiny packets of a chemical. The motor nerve impulse stimulates the motor nerve ending to release the chemical and make the muscle fibre contract. If a peripheral neuropathy affects the motor nerves, the muscles become weak because they do not receive the messages to move.

The cell body of a sensory nerve has two axons. One goes into the spinal cord and delivers messages to the brain. The other transmits messages from specialised receptors in the skin, joints and muscles. The receptors sense changes in pressure, position, or temperature or pain and translate the stimulus into a nerve impulse, which is relayed to the brain by the sensory nerve fibres. The fastest conducting nerve fibres are like telephone wires and have their own insulating sheaths. The sheaths are made of myelin, a fatty substance made by special cells, called Schwann cells. Nerve fibres conduct nerve impulses very quickly because the myelin sheath has gaps about every millimetre, which allow the nerve impulse to jump from gap to gap and travel faster. These fast-conducting myelinated nerve fibres control rapid movement and allow fine touch discrimination. There are also many nerve fibres without myelin sheaths. These are called unmyelinated fibres and conduct nerve impulses more slowly. They signal pain and temperature and are important for the control of blood circulation and sweating.



Different types of peripheral neuropathy

Most types of peripheral neuropathy usually come on very slowly over several months or years, a clinical course called chronic. Sometimes a peripheral neuropathy comes on very rapidly over the course of a few days, which is called acute. Intermediate courses, about four to eight weeks, are called subacute.

A peripheral neuropathy often affects all the nerves more or less together. Because the longest nerves are the most vulnerable, the feet and then the hands are most affected. Such a symmetrical pattern, affecting the feet and hands more than the hips and shoulders, is called a symmetrical polyneuropathy (*poly* means many). If only one nerve is affected, the condition is called a mononeuropathy (*mono* means single). If several discrete nerves are affected, the condition is called a multiple mononeuropathy (the term 'mononeuritis multiplex' might also be used). Sometimes the nerve roots (the name for parts of the nerves next to the spinal cord) are also affected, which gives rise to a polyradiculoneuropathy (*radiculo* means root). Polyradiculoneuropathy occurs in the common form of Guillain-Barré syndrome (GBS) and in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP).

A peripheral neuropathy usually affects sensory and motor nerve fibres together so as to cause a mixed sensory and motor neuropathy. Sometimes the autonomic nerve fibres are also affected. These control sweating, pulse, blood pressure, bladder, sexual and bowel function which may become affected. Sometimes a peripheral neuropathy just affects sensory nerve fibres, causing a pure sensory neuropathy. Finally the motor nerve fibres may be affected on their own, producing a pure motor neuropathy.

Nerve fibres may be damaged in four main ways:

- Most commonly, the delicate long axons lose their energy supply because of a chemical upset in the nerve cell body causing the axon to shrink. This is called an axonal neuropathy.
- Less commonly the problem lies in the insulating myelin sheath. This is called a demyelinating neuropathy.
- Vasculitis (inflammation of the blood vessels) may affect the nerves and cause a vasculitic neuropathy.
- Sometimes unusual chemicals or cells collect in the nerves and cause an infiltrative neuropathy.

Peripheral neuropathies do not affect the brain, vision, or the sense of smell. They almost never affect hearing and taste. Most sorts of peripheral neuropathy do not affect breathing or swallowing.

What is CIDP?

CIDP, or chronic inflammatory demyelinating poly-radiculo-neuropathy, is a rare inflammatory neuropathy that damages the peripheral nerves. ‘Chronic’ means ‘lasting a long time’. Although fewer people are newly diagnosed with CIDP each Around 3 people per 1,000,000 are diagnosed with CIDP each year. However, the prevalence, or number of people living with the condition at any one time, is around 5-7 per 100,000. CIDP is an autoimmune disease of the peripheral nerves, and symptoms experienced by patients can be similar to GBS, usually the symptoms of CIDP start more slowly.

CIDP is distinguished from GBS by timing. In GBS the worst point is reached within four weeks although may happen within a few days. By definition, CIDP continues to worsen more than eight weeks after onset. CIDP usually develops slowly over months, but sometimes a person initially diagnosed as GBS is later reclassified as ‘acute-onset CIDP’, because deterioration continues over an extended period, or several relapses occur after a period of improvement. In a large research study of patients initially diagnosed as GBS, 5% developed CIDP, and another 9% had a brief early relapse of GBS (‘treatment-related fluctuation’) but did not develop CIDP.

Occasionally, a patient who has recovered from GBS may develop GBS again many years later. This ‘recurrent GBS’ happens to around 3% of people with GBS. Sometimes a suspected recurrence of GBS is actually a relapsing-remitting form of CIDP.

Symptoms of CIDP

Symptoms of CIDP usually develop slowly starting in the feet and legs before progressing to other parts of the body. 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.

These symptoms may remain mild and result in only minor disruption of the patent’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.

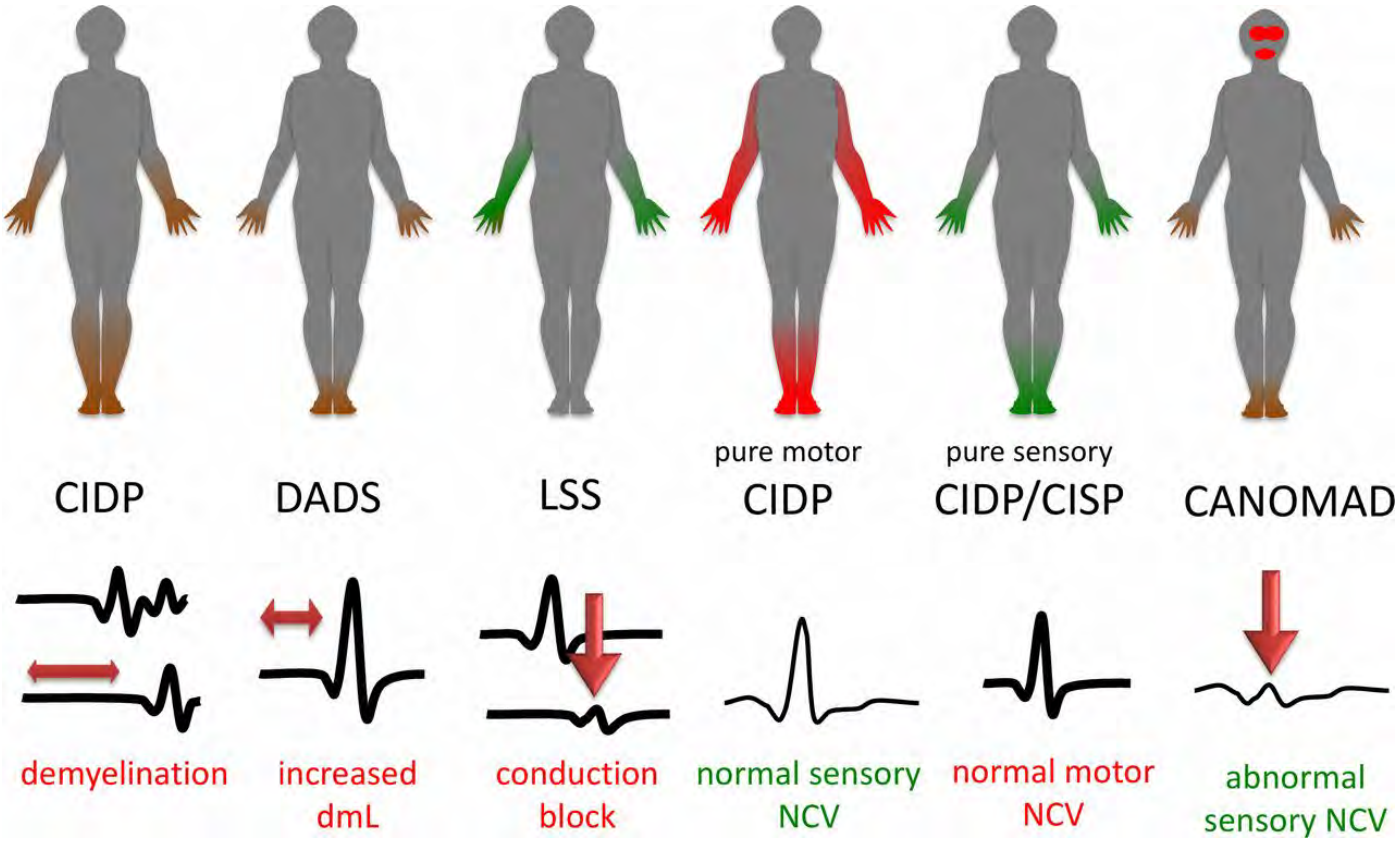
- numbness pins & needles
- muscle weakness
- fatigue
- loss of deep tendon reflexes

Acute onset CIDP (A-CIDP)

Due to similarity in the early stages up to 16% of patients will be misdiagnosed with GBS (AIDP), meaning patients may not receive the best treatment for their condition or understand the long-term outcomes.

Patients with A-CIDP are less likely to have autonomic nervous system involvement, facial weakness, a preceding infectious illness, or the need for mechanical ventilation. Around 8%-16% of GBS patients may relapse shortly after improvement or stabilization following initial immunological therapy. This is an important clinical issue because maintenance treatment is often required in CIDP. The diagnosis of A-CIDP should be considered when the condition of a patient with GBS deteriorates after nine weeks from onset, or when deterioration occurs three times or more.

Pattern of symptoms in chronic conditions



What causes CIDP?

Although sometimes a trigger is apparent, and in contrast to GBS, most patients with CIDP cannot identify a preceding viral or bacterial infection. GBS normally progresses over 2-4 weeks, then plateaus, improves over several months and does not usually

recur. CIDP has ongoing symptoms for over 8 weeks and usually does not improve unless ongoing treatment is given.

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 several 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 onset of symptoms
- 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

Nerve Tests

In hospital, two tests may be carried out to see how well your nerves are working. These are;

- electromyogram (EMG) – tiny needles are inserted into your muscles and electrical recordings are taken to see how they react when nearby nerves are activated.
- nerve conduction studies – small discs (electrodes) are stuck on your skin and minor electric shocks are used to activate the nerves and measure how quickly these signals travel along them.

In people with GBS, these tests will usually show that signals are not travelling along the nerves properly.

Lumbar Puncture

The lumbar puncture is a procedure to remove some fluid from around the spinal cord (the nerves running up the spine) which involves lying on one side and having a needle inserted into the base of the spine under local anesthetic.

The sample of fluid will be checked for signs of problems that can cause similar symptoms to GBS and CIDP, such as an infection.

Occasionally the diagnosis can be delayed for a few days while they are checking the results, and your doctor may start the treatment just to be on the safe side.

Treatment

The treatments with clinically proven efficacy in CIDP are immunoglobulin, steroids, and plasma exchange, all of which have been shown to be superior to placebo in clinical trials. In other studies, they have been shown to be equivalent to one another in efficacy: between 50 and 80% of patients respond to each of the treatments. Failure to respond to one treatment does not mean that the other treatments won't work either (unless the diagnosis if wrong), however, 15% of patients do not respond to any of the three treatments currently approved.

Immunoglobulin

The most commonly used treatment, particularly for motor dominant CIDP, is intravenous immunoglobulin (IVIg). Immunoglobulin is made from donated blood that contains healthy antibodies which can help stop the harmful antibodies damaging your nerves. If it works there will be a quick response in a matter of weeks, with improvement in strength and function. Some patients (15-30%) only need a single course.

IVIg is given in hospital and if it needs to be given long term by regular infusion typically every 6 weeks. This can have an impact on the patient's work, travel plans, or ability to look after family members, as their infusions need to be fitted around these factors. In some parts of the country it is possible to receive subcutaneous immunoglobulin (also known as SCIg or SubCut) at home which is more convenient for patients. 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.

As with all treatments, side effects can occur with IVIg, although usually these are minimal and do not require the treatment to be stopped. It is helpful if you drink plenty of fluid whilst you are receiving IVIg. Transient side effects, which often respond to changes in the rate of administration of the infusion, include headache and low blood pressure and occasionally, a rash can develop. Severe complications from IVIg use are very rare. IVIg thickens the blood slightly so particular consideration of its use is given to patients with kidney failure, previous heart disease, stroke or blood clots.

Plasma exchange (plasmapheresis)

Plasma exchange, also called plasmapheresis, is sometimes used instead of IVIg. This involves being attached to a machine that removes blood from a vein and filters out the harmful antibodies that are attacking your Plasma exchange involves having the patient attached to a machine to remove some of their blood and replace with other blood product (eg human albumin), over 5 days. PE should be considered in preference to IVIg in cases of non-motor dominant CIDP.

Corticosteroids

Corticosteroids should also be considered in preference to IVIg in cases of non-motor dominant CIDP. Corticosteroids are easy to administer either as daily or alternate daily treatments, or monthly oral or intravenous regimes. In older patients, there are often competing medical conditions which mean that they need to be used with caution or avoided altogether e.g. if a patient has poorly controlled diabetes or osteoporosis, or if they have recurrent chest infections and immunosuppression might put them at risk of

overwhelming infection. A younger patient may have fewer comorbidities but would also potentially be facing longer term treatment.

If a patient needs very frequent IVIg infusions then a neurologist might consider adding in a steroid or another immunosuppressant agent, and if IVIg does not offer clear benefits, then steroids are the obvious next treatment option. There is a difference in cost between the treatment options, with both IVIg and plasma exchange costing thousands of pounds per course whilst steroids are relatively cheap, but this is not a deciding factor in which treatment is provided.

Is there a cure for CIDP?

It depends how you define 'cure', but in CIDP literature this has been defined as 'remission of over 5 years off treatment', which has been reported in about 25% of patients treated with steroids for less than one year. Neurologists would more usually use the term 'remission' in CIDP (i.e. stability without treatment). Shorter term remission is more common than 'cure': there is evidence from studies that up to 40% of patients with CIDP may be in remission after one year of treatment with either steroids or IVIg. It is for this reason that it is advised that the need for treatment is reviewed on an annual basis if patients are no longer showing any or little fluctuation in their clinical symptoms. Although many people remain free of symptoms following remission, there is no guarantee that there will be no further relapse in the future. 15% of patients do not respond to any of the three treatments.

Other chronic conditions

Paraproteinaemic Demyelinating Neuropathy (PDN)

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)

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 is usually effective.

MADSAM also known as Lewis-Sumner syndrome

Multifocal acquired demyelinating sensory and motor neuropathy 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.

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, and CIDP is defined when the nadir comes after at least eight weeks. 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.

Chronic idiopathic axonal neuropathy

If no cause for the peripheral neuropathy can be discovered, doctors call it 'idiopathic' that means 'of its own cause'. This label probably covers a number of different causes which future research may uncover. With rare exceptions, chronic idiopathic axonal neuropathy occurs in older people, only worsens very slowly (and sometimes remains stationary), and does not become disabling. It is most commonly a sensory neuropathy causing numbness, tingling and discomfort in the feet that may gradually spread up the shins.

People may become slightly unsteady, and weakness of the ankles may develop. The amount of pain is variable. Some people have very little pain but more weakness. Others have little weakness but more pain.

Sensory neuronopathy

Also known as sensory ganglionopathy, dorsal root ganglionopathy or sensory ganglionopathy, this is a rare subgroup of peripheral nervous system diseases with specific characteristics, such as the primary and selective destruction of the dorsal root ganglia (DRG) neuron in the spinal cord and the trigeminal ganglia neuron in the skull.

It has a typical clinical presentation, with sensory deficits that are not dependent on length and patients often report a lack of coordination of muscle movements. There are several proposed mechanisms for the pathophysiology of the condition. The diagnosis is guided by the presenting symptoms and confirmed with diagnostic tests to differentiate from other related conditions.

Treatment of sensory ganglionopathy is often difficult and patients often stabilize but fail to improve significantly. There are reports of improvement in immune-mediated and paraneoplastic sensory ganglionopathy following treatment with intravenous immunoglobulin and other autoimmune medication, respectively. Quickly treating the underlying cause, such as the underlying neoplasia, may also be helpful.

Related Conditions

People that suffer from sensory neuronopathy are more likely to be affected by other autoimmune diseases. A differential diagnosis is important to distinguish sensory neuronopathy from other related conditions, such as sensory and ataxic neuropathy. These conditions may include Sjögren's Syndrome, autoimmune hepatitis and celiac disease.

POEMS syndrome

POEMS is a rare condition caused by the body producing abnormal plasma cells (a type of blood cell which produces antibodies to fight off infections). This blood disorder affects multiple organs in the body. It is named after the five common features of the syndrome described below.

Polyneuropathy - nerve damage leading to weakness, numbness and pain in the arms and legs

Organomegaly - an enlarged spleen and/or liver

Endocrinopathy - hormonal problems

M-protein or Monoclonal plasma cell disorder - an overproduction of abnormal plasma cells which lead to other multi system effects

Skin changes - darkening to skin, red spots on the body, hair growth

For more information, please see our POEMS fact sheet

Living with CIDP

Not everyone with CIDP will require a stay in hospital, but if CIDP comes on very rapidly, presenting more like GBS, it may be necessary to be admitted to ICU for a time. As this won't apply to them majority of people diagnosed with CIDP, more information about ICU is available on a separate information sheet.

If you do require a stay in hospital, you'll be closely monitored to check for any problems with your lungs, heart or other body functions. You'll also be given treatment to relieve your symptoms and reduce the risk of further problems. This may include:

- a ventilator if you're having difficulty breathing
- a feeding tube if you have swallowing problems
- painkillers if you're in pain
- being gently moved around on a regular basis to avoid bed sores and keep your joints healthy
- a thin tube called a catheter in your urethra (the tube that carries urine out of the body) if you have difficulty peeing
- laxatives if you have constipation
- medicine and/or special leg stockings to prevent blood clots

Once you start to improve, you may also need extra support to aid your recovery.

Depending on the severity of symptoms, and how well they respond to treatment, you may need help with everyday tasks, your home may need adaptations, or you may need a care package in place.

Introduce yourself to the doctor in charge of the case and don’t be afraid to ask questions. Get to know the regular nursing staff and ask for a daily update on progress.

Specialists who might support you through rehab:

physiotherapist	will help you with movement issues, re-building muscle, and learning to walk
occupational therapist (OT)	will identify potential problem areas, and work out some solutions
speech and language therapist	will help with communication and swallowing problems
counsellor or psychologist	will help you find ways to cope emotionally

Assessment & care plan

If you are likely to have ongoing health and social care needs you should have an assessment carried out by a multidisciplinary team of health or social care professionals such as a social worker, physiotherapist, occupational therapist, psychologist or dietician. You, and a family member if appropriate should be involved in this process.

A care plan should include details of:

the treatment and support you will get	who will provide the support	when and how often
monitoring and review	a named co-ordinator	who and how to contact

The type of support that might be in a care plan:

community care services	NHS continuing healthcare	NHS funded nursing care
rehabilitation	equipment	support from voluntary agencies

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 as described earlier.

Pregnancy

Neither IVIg nor plasma exchange is contraindicated during pregnancy. However, as plasma exchange requires additional considerations and monitoring, IVIg might be preferred.

Accessing physiotherapy

If you have difficulty accessing physiotherapy, but feel it would benefit you, or if you find yourself on a lengthy waiting list for community physiotherapy, please contact **GAIN** for access to our online physio videos free of charge.

Hygiene and cleanliness

Personal cleanliness for those who are unable to attend themselves fully can be a problem. You may have reduced use of your hands, maybe temporarily, but sometimes permanently. Some people may struggle to wash themselves, brush their hair, use the toilet independently, brush their teeth, cut their nails etc. It is important for both hygiene and self-esteem that these matters are attended to by a carer.

Dental hygiene

Through no fault of their own, many people's teeth are neglected during periods of serious illness. Once you have returned home, arrange an appointment with your dentist as soon as possible. If you have difficulty attending a dental practice, enquire about community dental services. Using an electric toothbrush can be helpful if you have weakness in your hands. More advice can be found on the British Society for Disability and Oral Health website; https://www.bsdh.org/index.php/how-to-find-a-special-care-dentist

Financial help

Financial concerns may be causing anxiety, especially if you are the main wage-earner and unable to work. If you are employed, stay in regular contact with employers and make sure you understand the absence and returning to work processes. There is more information about returning to work later in the booklet.

GAIN may be able to help with the cost of frequent journeys to visit a family member in hospital. Visit the website or get in touch and ask about our Personal Grants Scheme.

Once you leave hospital, or if you are already receiving treatment as an outpatient, you could be entitled to benefits to help you support yourself and your family. The benefits system is complex and subject to change, but Citizens Advice has up-to-date information on their website, and they can even help you make an application for benefits such as Personal Independence Payment (PIP) or Universal Credit: https://www.citizensadvice.org.uk/

Further information on benefits can be found at https://www.gov.uk/browse/benefits

If adaptations to your home are needed, such as installing a ramp or downstairs bathroom for example, it is worth contacting your local council to see if you are entitled to help via a Disabled Facilities Grant:

England and Wales
https://www.gov.uk/apply-disabled-facilities-grant
Scotland
http://www.disabilityscot.org.uk/info-guides/disabled-home-adaptations-1/
Northern Ireland
https://www.nidirect.gov.uk/articles/disabled-facilities-grants-0
Republic of Ireland
https://www.citizensinformation.ie/en/housing/housing_grants_and_schemes/housing_adaptation_grant_for_people_with_disability.html

If you are on a low income, or someone in your household is severely and permanently disabled, you may be entitled to a reduction in Council Tax. Further information is available in the Carers UK Council Tax factsheet: https://www.carersuk.org/images//Factsheets/2020-21_factsheets/Council_Tax_April_2020-21.pdf

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 if you need advice, ask to speak to a nutritionist. 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. Try to eat a healthy, balanced diet and keep consumption of sugary and processed foods, and drinks that are high in caffeine or alcohol to a minimum. There is some excellent advice on nutrition on the NHS website; https://www.nhs.uk/live-well/eat-well/

Your immune system

Although caused by your immune system malfunctioning, CIDP does not weaken or damage your immune system, and having CIDP does not mean that your immune system is compromised, unless you are being treated with steroids.

Regular exercise and maintaining a healthy diet and sleep regime will all help during recovery. There is anecdotal evidence, and lots of hype surrounding supplements and alternative remedies, but very few are known to have real benefits. Most people are able to get the vitamins they need from their diet, but if you wish to take dietary supplements, your doctor (rather than Google!) will be able to advise you on anything that may help you during recovery.

Exercise and rest

Physiotherapy, occupational therapy (OT) and speech and language therapy play a vital role in maximizing functional ability.

Exercise can help to improve your muscle strength and reduce your overall sense of fatigue. It can also aid sleep and support mood through the production of endorphins. Other benefits include helping your heart and lungs remain healthy and making you feel more positive about yourself. However, it may take weeks or even months before you feel some of the benefits, so it is important to pace yourself. You will find lots of exercises online, including seated exercises, or you could ask your GP or physiotherapist about how to start regular exercise and what exercises might be right for you.

Visit the NHS website for advice on exercise: https://www.nhs.uk/live-well/exercise/ and also for advice on sleep: https://www.nhs.uk/live-well/sleep-and-tiredness/

Pain

Being in pain naturally impacts on mood and the ability to cope with everyday situations. If you are experiencing pain, it is important to talk to your family and friends about this, so that they understand what you are going through and why you might be irritable and difficult to live with at times.

Some other sources of support are shown below.

Other sources of support	PAIN
Pain Concern	https://painconcern.org.uk/
British Pain Society	https://www.britishpainsociety.org/
Pain Association Scotland	https://painassociation.co.uk/
Welsh Pain Society	https://www.welshpainsociety.org.uk/
Pain Relief Foundation	https://painrelieffoundation.org.uk/
Pain Management NI	https://www.myni.life/pain-management
Pain Relief Ireland	https://painreliefireland.ie/

Mental health & wellbeing

With all the changes in your health it is not uncommon to feel anxious or angry, and this can even be helpful to a degree in providing the mental and physical energy that is needed to anticipate and tackle problems. An acute stress reaction is recognised as being a normal part of the process of adjusting to a life change, and it can often help to talk things through with a partner or close friend. However, please seek advice and help from your GP if you are finding it difficult to cope or to sleep, or if you are feeling overwhelmed emotionally.

The impact of being severely affected by a sudden, frightening and potentially life-changing condition such as CIDP can leave people with symptoms of PTSD (post-traumatic stress disorder). It can develop immediately, or many months or even years after a traumatic event. If you are concerned about low mood, irritability, panic attacks, anxiety, flashbacks, physical sensations such as trembling or sweating, poor concentration or sleeping problems, please talk to your GP about getting some counselling.

Some other sources of support are shown below.

Other sources of support	MENTAL HEALTH
5 steps to mental wellbeing	https://www.nhs.uk/conditions/stress-anxiety-depression/improve-mental-wellbeing/
Mindfulness	https://bemindful.co.uk/
Post-Traumatic Stress Disorder	https://www.nhs.uk/conditions/post-traumatic-stress-disorder-ptsd/
Mental health and wellbeing	https://www.nhs.uk/conditions/stress-anxiety-depression/
MIND	http://www.mind.org.uk/
Young Minds	https://youngminds.org.uk/

Sexual relationships

CIDP and other chronic neuropathies can bring on problems in any relationship, and 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. As well as coming to terms with a 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 relationship support

Relate: https://www.relate.org.uk/

Sexual Advice Association: https://sexualadviceassociation.co.uk/

Getting back to work

When someone is ready to return to work after an absence, the employer should have a procedure they follow. You can check your workplace's absence policy for this. Your workplace might have a policy for meeting with employees after absences.

After long-term absence

If there's been a long absence or the employee has an ongoing health condition, it's a good idea for the employer and employee to meet and:

- make sure the employee is ready to return to work
- talk about any work updates that happened while they were off
- look at any recommendations from the employee's doctor
- see if they need any support
- if the employee has a disability, see if changes are needed in the workplace to remove or reduce any disadvantages ('reasonable adjustments')
- consider a referral to a medical service such as occupational health
- discuss an employee assistance programme (EAP) if it's available
- agree on a plan that suits you both, for example a phased return to work

Making reasonable adjustments

If an employee has a disability, by law their employer must consider making 'reasonable adjustments' if needed to help them return to work.

Reasonable adjustments could include making changes to the employee's:

- workstation or working equipment
- working hours
- duties or tasks

This can help:

- get people back to work quicker
- prevent any further problems

For the best course of action, the employer should take advice from:

- the employee themself
- the employee's doctor
- their occupational health adviser

Phased return to work

A ‘phased return to work’ is when someone who has had a long-term absence might need to come back to work on:

- reduced hours
- lighter duties
- different duties

The employer or their HR manager and the employee should agree on a plan for how long this will be for. For example, they could agree to review how things are going after a month and then decide to increase the working hours or duties, or they might decide they need to stay reduced for longer.

The employer or HR manager should continue to regularly review the employee’s health and wellbeing in the workplace and make new adjustments if necessary.

Example of a phased return to work

Stage	Total days worked in a week	No. of days worked consecutively	No. of hours worked in a week	No. of hours worked per day				
				Mon	Tue	Wed	Thu	Fri
1	1	1	3-5	ANY DAY				
2	2	1	6-10	--	4	--	4	--
3	3	1	12-15	4.5	--	4.5	x	4.5
4	4	2	20	5	5	--	5	5
5	4	2	24	6	6	--	6	6
6	4	2	30	7.5	7.5	--	7.5	7.5
7	4	3	30	7.5	7.5	7.5	--	7.5
8	4	4	30	7.5	7.5	7.5	7.5	--
Last	5	5	37.5	7.5	7.5	7.5	7.5	7.5

Pay during a phased return to work

If the employee returns to their normal duties but on reduced hours, they should get their normal rate of pay for those hours they work. For the time they're not able to work, they should get sick pay if they're entitled to it.

If the employee is doing different duties, it's up to the employer and employee to agree on a rate of pay. It’s a good idea to make sure this agreement is put in writing.

This information is provided by ACAS (the Advisory, Conciliation & Arbitration Service). Further information regarding absence and returning to work is available on the ACAS website: https://www.acas.org.uk/absence-from-work

Vaccinations

Having had GBS, or living with CIDP does not mean you are at increased risk from vaccines. All vaccines available in the UK are safe and reports made to the Yellow Card Scheme from the many millions of COVID vaccinations administered, would indicate there are no concerns of serious side effects.

Millions of people, including people who are living with CIDP, have a flu jab every year with no problem. It remains the best protection against flu and the very serious risk it can pose to the most vulnerable. It is recommended that people with a chronic neurological condition such as CIDP protect themselves from respiratory viruses such as flu, COVID, etc. by being vaccinated.

Although studies over the last couple of decades have found there may be approximately one extra case of GBS/CIDP per 1,000,000 flu immunisations, the view of both Public Health England and the MHRA is that there is insufficient evidence to point to a definite causal link. Whatever link there may be represents a very small risk in comparison to the much greater risk posed by viruses such as flu and COVID, both in terms of the danger of the virus itself to people most at risk, and also the risk of the flu virus triggering GBS/CIDP (COVID does not appear to be a significant trigger for GBS, if at all).

The vast majority of people will have no serious side effects following a vaccination, and with respect to COVID-19, vaccines are our only chance to achieve herd immunity, protect the most vulnerable amongst us and return to normality. The advice of our Medical Advisory Board therefore is that most people, especially those considered at greatest risk from flu and COVID, should be vaccinated.

None of the COVID vaccines approved to date is live attenuated, but should a live vaccine become available, then this should be avoided by anyone whose immune system is suppressed, due to taking corticosteroids to treat CIDP for example.

On vaccinations in general

On vaccinations in general, our Medical Advisory Board advises:

- DON'T have unnecessary vaccines for travel but DO have all travel vaccines that are recommended for the particular area you are travelling to.
- DO have all vaccines that are 'necessary'. This includes the flu vaccine (if you are in an at risk group), MMR, DTP, HIF, COVID-19, etc. T**here are monitoring programmes ongoing so a link would be picked up if it occurred.**
- COVID-19 is a more serious disease than influenza and more easily caught. Most people with GBS or CIDP should receive any of the COVID-19 vaccines, except perhaps people with a history of severe allergy requiring an Epipen.

Having relatively mild side effects such as numbness and tingling is quite common following a vaccination, and is almost certainly nothing to be concerned about. If you have a chronic neuropathy such as CIDP, a vaccination might cause a slight 'flare-up' of symptoms due to your immune system being stimulated. Most will only last a few days, but if they last longer than this, or if symptoms get worse or start spreading, then consider contacting your GP. Anyone can report side effects of medication or vaccines, regardless of severity, and if you would like to do so, please follow this link; https://coronavirus-yellowcard.mhra.gov.uk/

Vaccines currently in use are amongst the safest medicines available. However, as with many things in life, there is no simple 'yes or no' answer, and each person must weigh up the risks associated with not having a vaccination, against the very small risk of an adverse reaction that might be associated with having it. Hopefully, this information will help you reach an informed decision.

Advice for carers

Hospital discharge can bring mixed emotions. Of course, you're glad to have your family member home, but it is quite normal to feel a bit daunted by what lies ahead, especially if you are now the main carer. Here are a few practical suggestions that can help you in your new role. Other sources of support are listed overleaf.

Ask for a needs assessment for the person you're caring for	You need this before the council can recommend services such as equipment, home adaptations, help from a paid carer, etc (see table overleaf)
Get a carer's assessment	This can be requested at the same time as a needs assessment, via social services at your local council (see table overleaf)
Don't be afraid to ask for help	Ask family and friends (shopping, respite, etc), and contact carers' organisations or Citizens Advice for benefits advice and other support (see table overleaf)
Look after yourself	Eat a healthy diet, maintain a regular sleep pattern and make time for exercise, preferably in the fresh air or at your local gym or pool, away from the care setting
Remember to rest	Put some activities on hold. Take some time for yourself by arranging respite care from a friend or relative, or through social services
Recognise limitations	Both those of the person you're caring for and your own. Let them try to do things for themselves if they are able, but bear in mind their safety and yours
Look after your mental health	Talk to each other about your experiences and frustrations during illness and recovery. Talk to your GP if either of you is struggling
Be mindful of risks	Be aware of temperature (cooker, iron, hot bath water, hot drinks), risk of falls due to poor balance, and dropping or spilling things due to weakness
Diet & exercise	If appetite and taste are affected, ask about vitamin supplements to help maintain dietary needs. Ask the community physio about exercises to do between visits
Don't lose sight of your relationship	Whatever your relationship is to the person you're caring for, make time for it. Do things together that you both enjoy and that feel 'normal'
Get in touch with GAIN	We provide information and support, organise peer support via phone or video link, virtual group chats, and run an active and supportive Facebook group

Looking after someone you love doesn't have to mean struggling alone. There are organisations out there who offer advice and practical support to carers, so reach out and see what help is available to you.

Other sources of support	CARERS
NHS advice	https://www.nhs.uk/conditions/social-care-and-support-guide/support-and-benefits-for-carers/
Carers UK	https://www.carersuk.org/
Carers Scotland	https://www.carersuk.org/scotland
Carers Wales	https://www.carersuk.org/wales
Carers Northern Ireland	https://www.carersuk.org/northernireland
Family Carers Ireland	https://familycarers.ie/
Carers Trust	https://carers.org/
Help for young carers	https://youngminds.org.uk/find-help/looking-after-yourself/young-carers/
Citizens Advice UK	https://www.citizensadvice.org.uk/
Citizens Information RoI	https://www.citizensinformation.ie/en/
Find your local social services	England https://www.nhs.uk/service-search/other-services/ Scotland https://www.mygov.scot/find-your-local-council/ Wales https://111.wales.nhs.uk/localservices/ Northern Ireland http://online.hscni.net/social-services/ Republic of Ireland https://www.hse.ie/eng/services/list/4/

My child has been diagnosed with CIDP

Although more common in adults, CIDP can affect anyone of any age. It is distressing for any parent to see their child debilitated by illness, and we hope that knowing a little bit about what to expect may help, especially in the early stages following diagnosis. Don't forget your child needs to be told what is happening and will be reassured by familiar faces and voices.

If your child is of primary school age, ask us for an Activity Pack containing games and stories designed to can help your child understand what is happening. It's free of charge, and even comes with a cuddly tortoise! If you would like to talk to another parent of a child affected by CIDP, please contact us by email or by phone, and we will arrange contact with one of our support volunteers.

How is my child's condition managed?

Diagnosis and treatment will follow a similar path for children as for adults, and are covered elsewhere in this booklet. Paediatric services tend to be very self-contained, and you will have access to support that you would not experience in an adult setting. The following information is specific to differences between adult and paediatric services. It is important your child is treated in a centre familiar with CIDP and other inflammatory neuropathies, so they may be moved to a hospital you do not routinely use.

People likely to be involved in your child's care:

- you, your child and your family
- paediatric nurses and nurse practitioners, doctors and neurologists
- paediatric intensive care nurses and doctors
- neurophysiologist, who does the nerve conduction study
- physiotherapists, to help with movement and breathing
- speech therapist, to help with feeding and communication
- occupational therapist, to help maximise recovery
- clinical psychologist

Other staff you may meet could include dietician, health care assistants, family liaison nurses, play specialists, chaplains and interpreters.

Things you can do to help

- **Talk about things that matter** to your child (a pet, the football results, family events, messages from friends, etc).
- **Maintain the general routine as much as possible.** It's important to communicate to the patient and their siblings that life goes on, that this illness is a bump in the road and not a dead end.
- **Maintain rules and responsibilities.** Do not excuse a child recovering from CIDP from responsibilities if they are capable. Revise expectations, but being a part of a family means helping in some way. A recovering child maintains a sense of belonging and usefulness when he or she can contribute.
- **Answer questions based on fact.** Young children can make up reasons for the illness that exist in the magical realm. Children may believe that their comments or actions can be connected to the cause of the disease. That said, the reality of the disease will bring up feelings and questions to be addressed. Every feeling that a child has should be discussed and acknowledged.
- **Involve siblings in the recovery process.** Siblings can help with homework, physical therapy, and making accommodations for the patient. Be careful not to rely too much on siblings – their experience of childhood is equally important.
- **Allow the patient to participate in medical decisions.** Giving choices to a child develops responsibility and reasoning skills. If a child can handle it, participating in treatment choices can be extremely beneficial to their emotional development. Scheduling decisions, describing physical sensations to doctors and nurses, learning to adjust to social interactions, learning to communicate their limitations to friends, and making accommodations in the home and at school – all of these can be a discussion with the patient, giving them a sense of power in a powerless situation.

Rehabilitation and outcome

The majority of children respond well to treatment, and around 80% go on to make a full recovery, albeit this can be a long and sometimes painful process.

Keep in touch with school, and once your child is ready, ask their teachers for any resources they can access to help them catch up with what they would have been doing at school.

Going home

Occupational therapists will discuss the suitability of the home environment and tell you how to access any specialised equipment that might be needed.

Hydrotherapy and swimming can be very beneficial, as the water is weight-bearing allowing freer movement. Consider going for a family bike ride once your child is strong enough, and even look at an activity such as horse-riding, as this can help with balance as well as the psychological benefits associated with being around animals (contact 'Riding for the Disabled', https://www.rda.org.uk/). Exercise is much easier when children are engaged, so it helps a lot if it is something fun and enjoyable. Since children are usually very active and move around without thinking, observing what they do naturally is a good indicator of their capabilities.

Your child may feel the cold and get tired more easily, or they may lose their appetite or complain of things tasting different. CIDP may also have an effect on their mood or behaviour; they may become frustrated, angry and upset at not being able to do everything that their friends can, and that they could do before. All of this is normal. It is important to listen to them, understand what they are feeling, and try to find things that will help. Be patient. These problems are temporary and will improve with time, but talk to your GP if you are concerned and feel further support is needed.

Back to school

Thousands of children live with health issues that involve long periods of absence from school. At some stage, these children will face the unenviable task of going back to school and reintegrating into their former class and its usual daily routines. Catching up with work and friendships can often be difficult, but a good school will do all it can to ease your child's passage back into school life.

Returning to school: catching up with work

When your child is ready to return to school, the thought of catching up on weeks of missed work can be daunting. But, says Lesley Black, special educational needs (SEN) advisor at the charity Contact, ‘no school will want to overwhelm a child who has been unwell or dealing with difficult circumstances by expecting them to do extra homework or catch up on work during playtimes.’

Schools may prioritise catching up with work in maths and English – the core subjects – over other subjects. It’s quite common for schools to have small groups of children who need support with numeracy or literacy, and children who’ve been away from school will often slot into one of these. They may be given extra one-to-one help by a teaching assistant (TA) to help them catch up.

If you want to give your child a boost at home, speak to their teacher about how you can best help, such as games-based learning activities online that consolidate maths and English skills, or practical things you can do like reading aloud and cooking together. However, be aware that if your child has been ill they might find the school day very tiring, and may not be up to doing much homework.

Returning to school: fitting back in

Despite the fact that schools are under increasing pressure to ensure children achieve, your child’s school is likely to be far more concerned about their wellbeing as they return to school. Your school should help your child to keep in touch with their classmates while they’re absent, for example through visits, emails, letters or video calls. They also have a duty to help your child reintegrate once they’re well enough for school.

Your child may feel awkward if they’re bombarded with questions about why they’ve been away. Sometimes, they’re so reluctant to draw attention to themselves that they put up with discomfort or distress rather than asking a teacher for help. Some children, however, are happier with everything out in the open and even like to stand up in front of the class to explain what’s been happening. If your child has a specialist nurse or a social worker, they may be happy to visit the class, either with or without your child, to answer their classmates’ questions.

‘It’s important that schools are sensitive about your child’s needs when they’re considering what information to share with their classmates and the wider school community, and that they make sure your child has someone they can ask for help,’ Lesley says. ‘This could be a school counsellor or the special educational needs co-ordinator (SENCO) rather than their class teacher.’

Returning to school: the Individual Healthcare Plan

A key part of your child’s return to school after illness may be the creation of an Individual Healthcare Plan. This is a formal document that sets out things like:

- What their condition is

- What medication they take
- Who, in the school community, can administer their medication
- What to do in an emergency
- Arrangements that need to be made to enable your child to attend school, such as a quiet rest area

'You and your child should be central in drawing up the individual healthcare plan,' says Lesley. Other people involved in your child's care may also be involved, such as their GP, paediatrician or specialist nurse. The plan will identify if anyone needs to be given special training to administer medication or otherwise help care for your child at school. If so, someone from your child's medical team is likely to arrange this. The plan should be reviewed regularly, and at least once a year.

Depending on the level of recovery or any residual disability, it may be necessary for your child to return to school in a wheelchair, and school facilities should be checked to make they are suitable for your child's needs. A welfare officer from the local education authority (LEA) can visit to arrange adaptations, and can sometimes provide equipment for use in schools. Like any other institution, the ability of the school to cope depends on the willingness of those involved, as much as the building's design and facilities.

Before your child returns to school, arrange a consultation with headteacher and SENCO to explain what if any limitations they have.

This information has been adapted from TheSchoolRun: an online resource for parents of children at primary school:
https://www.theschoolrun.com/catching-up-at-school-after-absence

More detailed information is published online by Contact: https://contact.org.uk/advice-and-support/education-learning/attendance-absence-medical-needs/help-at-school-if-your-child-has-medical-needs/

Can you tell me more about GBS?

CIDP, or chronic inflammatory demyelinating polyneuropathy, is less common than AIDP (the acute version of the syndrome usually referred to as GBS). Although fewer people are diagnosed with CIDP each year (around 1-2 people per 200,000), the prevalence, or number of people living with the condition at any one time, is estimated to be 5-7 per 100,000. Like GBS, CIDP is an autoimmune disease of the peripheral nerves, and symptoms experienced by patients with both conditions can be very similar, although in most cases, the symptoms of CIDP develop more slowly.

CIDP is distinguished from acute GBS by virtue of its pattern of progression. GBS is defined if the worst point is reached within four to six weeks although this might happen within a few days. The initial progressive phase of CIDP usually lasts longer, but occasionally presents as acute-onset CIDP, and is initially diagnosed as GBS. It is only when the deterioration continues over an extended period, or when one or more relapses occur after a period of improvement, that the condition is reclassified as CIDP.

Occasionally, a patient who has recovered from GBS experiences similar symptoms several months or even years after recovery, and might be diagnosed with a recurrence of GBS. A recurrence is rare, and consideration should be given to the possibility that it may in fact be a slowly progressing or remitting and relapsing form of CIDP.

If you wish to know more about CIDP and the chronic variants, please visit our website or see our information booklet of the same name.

Contact GAIN if you or a member of your family has received a diagnosis of:

ACUTE

GBS Guillain-Barré syndrome, also known as

AIDP Acute Inflammatory Demyelinating Polyneuropathy

MFS Miller Fisher syndrome

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

AMSAN Acute Motor Sensory Axonal Neuropathy

BBE Bickerstaff's Brainstem Encephalitis

Brachial Neuritis also known as **Neuralgic Amyotrophy**

SIDP Sub-acute Inflammatory Demyelinating Polyneuropathy

CHRONIC

CIDP Chronic Inflammatory Demyelinating Polyneuropathy

A-CIDP Acute-onset CIDP

CIAN Chronic Idiopathic Axonal Neuropathy

CMFS Chronic Miller Fisher syndrome

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

DADS Distal Acquired Demyelinating Symmetric neuropathy

MMN/CB Multifocal Motor Neuropathy/with conduction block

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

PDN Paraproteinaenemic Demyelinating Neuropathy

sometimes described as CIDP with **Paraproteinemia**

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

Sensory Neuronopathy also known as **Sensory Ganglionopathy**