

The History of Guillain-Barré Syndrome

Landry's Ascending Paralysis

Descriptions of progressive numbness and weakness over a short period followed by spontaneous recovery exist in medical papers of the early 19th century. However the peripheral nervous system was little understood so no informed explanation for the symptoms was possible. It was not until 1848 (Graves) that it was suggested that such paralysis came from within the nervous system

Certainly the best description of “ascending paralysis” in this period was made by a Frenchman named Jean Baptiste Octave Landry de Thezillat in 1859.

Landry's description was based on ten cases, five of his own and five from the medical literature. In one case, Landry gave a long description of a patient who eventually died of asphyxia. Landry's superior, a Dr Glauber who had admitted the patient, had diagnosed hysteria but Landry predicted the patient's demise at an early stage.

Landry offered no explanation as no abnormality was found during a post mortem. Glauber added a note however, speculating a close connection between Landry's cases and the paralysis that follows diphtheria.

While Guillain-Barré syndrome was subsequently found to be unconnected with diphtheria, Glauber's observation was vindicated as both types of paralysis are due to a demyelinating neuropathy.

A translation of Landry's paper reads:

The sensory and motor systems may be equally affected. However the main problem is usually a motor disorder characterised by a gradual diminution of muscular strength with flaccid limbs and without contractures, convulsions or reflex movements of any kind. In almost all cases micturition and defecation remain normal. One does not observe any symptoms referable to the central nervous system, spinal pain or tenderness, headache or delirium. The intellectual faculties are preserved until the end. The onset of the paralysis can be preceded by a general feeling of weakness, pins and needles and even slight cramps Alternatively the illness may begin suddenly and end unexpectedly In both cases the weakness spreads rapidly from the lower to the upper parts of the body with a universal tendency to become generalised.

The first symptoms always affect the extremities of the limbs and the lower limbs particularly. When the whole body becomes affected the order of progression is more or less constant: (1) toe and foot muscles, then the hamstrings and glutei, and finally the anterior and adductor muscles of the thigh; (2) finger and hand, arm and then shoulder muscles; (3) trunk muscles; and

(4) respiratory muscles, tongue, pharynx, esophagus, etc. The paralysis then becomes generalised but more severe in the distal parts of the extremities. The progression can be more or less rapid. It was eight days in one and fifteen days in another case which I believe can be classified as acute. More often it is scarcely two or three days and sometimes only a few hours.

When the paralysis reaches its maximum intensity the danger of asphyxia is always imminent. However in eight out of ten cases death was avoided either by skillful Professional intervention or a spontaneous remission of this phase of the illness. In two cases death occurred at this stage. When the paralysis recedes it demonstrates the reverse of the phenomenon which signaled its development The upper parts of the body, the last to be affected, are the first to recover their mobility which then returns from above downwards.'

The term "Landry's ascending paralysis" was first used in 1876 (Westpahl). The usual treatment was with strychnine which probably did the unfortunate patients more harm than good. Landry contributed no more to neurology for he died of cholera just six years after publishing his paper.

Acute Febrile Polyneuritis

Acute febrile polyneuritis was one of six classes of polyneuropathy proposed by Ostler in 1892. Ostler considered that some of Landry's patients had fallen into this category while others had suffered from myelitis (inflammation of the myelin).

Ostler's description was of an illness similar to what we now call GBS but with the fundamental difference of showing a fever. In 1918, Bradford et al described 'acute infective polyneuritis'. They stated this to be the same as acute febrile polyneuritis, any fever having recovered before the onset of the neurological symptoms or being due to subsequent infection.

A Syndrome of Radiculoneuritis...

Guillain and Barré were medical students together at the Saltpetriere in Paris at the turn of the century and specialised in neurology. During the First World War, they were both serving as doctors in the French Army. They noted the cases of two soldiers who had become partially paralyzed. One, in particular, had fallen over when he had put his pack on and had been unable to get up. Both the soldiers quickly recovered, possibly assisted by treatment with pork chops and claret.

Together with Strohl, they published their classic paper in 1916. It was noted that reflexes were reduced and that the protein level in the cerebrospinal fluid was raised though this was not accompanied with a high white blood cell count. This was a crucial discovery as two common infections of the time, syphilis and tuberculosis, would have shown such an increase.

The cause of the condition was left unanswered, assumed to be some unknown kind of **GBS** infection or poisoning. Guillain personally was not convinced that the condition that he and his partners had described was the same as Landry's. Landry had noted how the condition could

cause respiratory failure but Guillain saw no such evidence and believed the illness he had described was not particularly serious.

Guillain Barré Syndrome

After World War One, doctors were faced with three similar conditions with slightly different definitions: Landry's ascending paralysis, acute febrile (or perhaps infectious) neuropathy and the radiculoneuritis described by Guillain et al.

It was in 1927 when the term Guillain-Barré syndrome was first used at a presentation by Dragonescu and Claudian. Their presentation was introduced by Barré himself but Strohl's name was omitted not only from the title of the presentation but also from the list of authors in the reference to the 1916 paper.

Later, and inevitably, it was suggested that the three conditions were one but Guillain would not countenance it. He emphasized that fever was not in his description, that the essential element of his definition was the raised cerebrospinal protein level which Ostler and others had not mentioned (because lumbar punctures had not been introduced), and that Landry's cases were a miscellany of conditions including poliomyelitis and encephalomyelitis.

In 1949 Haymaker and Kernohan suggested a wider definition of the illness, suggesting that Landry's ascending paralysis and Guillain-Barré syndrome were indistinguishable and called the condition Landry-Guillain-Barré syndrome. Guillain, who two years previously had retired from his position of Professor of Neurology at the Saltpetriere, was outraged and continued to stress his own narrower definition.

Guillain's last paper was in 1953. He believed the syndrome to be *generally* benign though the death of a patient, who after postmortem was found to have extensive peripheral nerve damage, had necessitated a shift in his position. Guillain still suspected an unknown infection as the cause and dismissed suggestions, made ten years earlier (Bannwarth), that the cause was due to allergy.

Later Developments

In 1956, C. Miller Fisher, a US doctor, described three patients with acute external ophthalmoplegia (eye paralysis), sluggish pupil reflexes, ataxia (lack of balance) and areflexia (absent tendon reflexes). Two patients had no weakness; the other had a facial palsy and possible weakness. All three recovered spontaneously.

Because some patients with GBS had ophthalmoplegia and there were other similarities, Dr Fisher concluded that these patients had suffered a disorder akin to GBS.

In 1958, a paper was published by Dr JH Austin who described a chronic form of GBS. Austin's paper was based on a review of 30 cases, the earliest of which went back to 1894, and on two of his own. This chronic form has gone through a variety of names and attempts to define it though it now generally known as CIDP.

Guillain died in 1961 and Barré in 1967. Having first published the relationship between ascending paralysis and an increased protein count in cerebrospinal fluid in 1916, they had seen a huge increase in knowledge as well as witnessed the use of early intensive care techniques.

So while Guillain and Barré continue to receive the recognition, spare a thought for poor neglected Strohl who history has ignored. Consider too the work of Landry, the victim of an early death from an illness he contracted from his own patients.

Information

A Quick Guide to GBS and CIDP

What is GBS?

GBS is short for 'Guillain-Barre syndrome' (pronounced Ghee-lan Bar-ray). It is an acute disease of the peripheral nervous system in which the nerves in the arms and legs become inflamed and stop working. This causes sudden weakness leading to limb paralysis, and a loss of sensation, sometimes with pain.

What is CIDP?

Some patients have a similar but longer-lasting illness called CIDP (chronic inflammatory demyelinating poly [radiculo] neuropathy). CIDP, once known as 'chronic GBS', is now usually regarded as a related condition.

Who can get GBS and CIDP?

Anyone: young or old, male or female. The illnesses are neither hereditary nor contagious. GBS affects about 1500 people every year in the United Kingdom; the incidence of CIDP is perhaps one tenth that of GBS.

What causes GBS/CIDP?

This is a matter of much research. About sixty percent of patients suffer from a throat or intestinal infection, influenza or stress symptoms in the previous two weeks. These infections trigger an incorrect response in the immune system which attacks the nerves.

What are the symptoms?

First symptoms are usually tingling and numbness in the fingers and toes with progressive weakness in the arms and legs during the next few days. In the mildest of cases, the weakness may arrest and cause only moderate difficulty in walking, requiring sticks, crutches or a walking frame.

In some cases the weakness progresses and leads to complete paralysis of the legs, the arms may also be affected. In a quarter of cases the paralysis progresses up the chest and the patient is unable to breathe on his or her own and needs to rely on a mechanical breathing machine (ventilator). The throat and face may be affected making swallowing impossible and so the patient needs to be fed by tube up the nose or directly into the stomach.

Occupational therapy

If you have problems with activities involving your hands, ask whether a referral to an occupational therapist would be helpful. Occupational therapists may be able to recommend arms may also be affected. In a quarter of cases the paralysis progresses up the chest and the patient is unable to breathe on his or her own and needs to rely on a mechanical breathing machine (ventilator). The throat and face may be affected making swallowing impossible and so the patient needs to be fed by tube up the nose or directly into the stomach.

For CIDP patients, the illness follows a longer course but respiratory failure is highly unlikely.

How are GBS and CIDP diagnosed?

From the history and clinical examination. This is difficult because the symptoms may be confused with those of other conditions.

Two confirmatory tests may be helpful and are performed in most cases:

- lumbar puncture - under a local anesthetic, a needle is inserted between the lower back bones and a small amount of spinal fluid is drawn off for analysis; and
- electromyogram (EMG) - an electrical recording of nerve conduction and muscle activity.

What is the treatment for GBS?

GBS improves spontaneously. However, certain factors can assist recovery:

- good nursing and medical/intensive care,
- physiotherapy and hydrotherapy, therapies that relieve discomfort and prevent stiffness,
- plasmapheresis - the exchange of blood plasma generally reduces the duration of the disease in severe cases if carried out in the first few days,
- immunoglobulin - the infusion of immunoglobulin proves successful with similar results to Plasmapheresis, and
- counseling to reassure the patient and encourage the patient towards recovery.

What is the treatment for CIDP?

Like GBS, CIDP can improve without treatment. However, recovery may be very slow and the illness can either get progressively better or worse, or can follow a relapsing/remitting course.

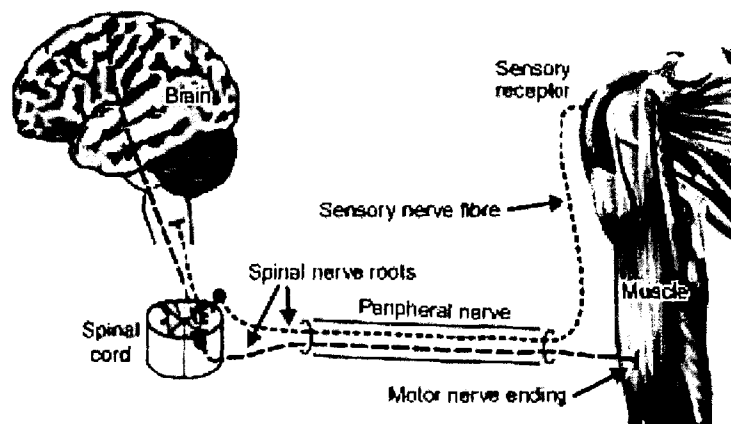
GUIDE

Peripheral Nerve Disorders

Introduction

How peripheral nerves work

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 in 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.

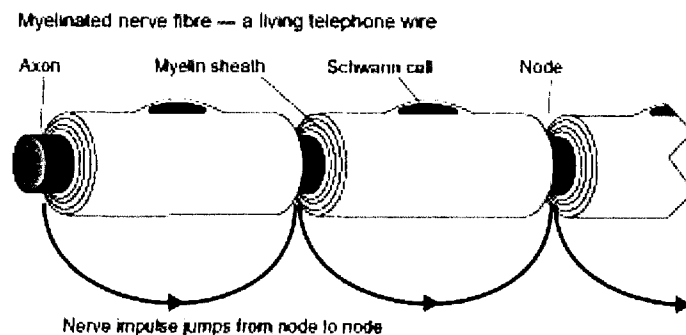


The motor nerve cell body has a long fibre called an axon, which extends from the central nervous system to the muscles. The longest axons can be as much as a metre long, for example the nerves to the muscles in the feet. The connection between the motor axon and the muscle fibre is a specialised nerve 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 goes out to specialised receptors in the skin, joints and muscles. The receptors sense changes in pressure, position, or temperature or pain. The receptor translates the stimulus into a nerve impulse. The sensory nerve fibres relay the impulses to the brain.

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 unmyelinated fibres 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 old-fashioned term 'mononeuritis multiplex' is also used). Sometimes the nerve roots (the name for parts of the nerves next to the spinal cord) are affected as well, which gives rise to a **polyradiculoneuropathy** (radiculo- means root). Polyradiculoneuropathy occurs in the common form of Guillain-Barré syndrome and in chronic inflammatory demyelinating polyradiculoneuropathy.

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.

Symptoms of a peripheral neuropathy

A peripheral neuropathy may be very mild. **Many people do not have any symptoms at all** but are discovered to have a peripheral neuropathy when they have a medical examination. The doctor may find signs of such mild neuropathies during a routine medical examination.

The first symptoms of a symmetrical neuropathy are usually very slight loss of feeling together with pins and needles in the toes and the soles of the feet, like an anaesthetic wearing off or like the feeling after having crossed your legs for too long. Some patients cannot feel their feet, others feel as though they are wearing socks or have cold feet. If the peripheral neuropathy worsens, similar feelings may affect the fingers. Sometimes a peripheral neuropathy is painful. The pain is often pricking or stabbing and made worse

by touching. It may also be aching or burning. Strangely an area, which is numb, may be painful or even supersensitive, so that a slight touch, which would not normally hurt, feels very unpleasant.

If the motor nerve fibres are affected, weakness may occur. This may cause difficulty running or walking fast. The toes may tend to catch in pavements. Slight unsteadiness may become a problem, especially in the dark or on rough ground. In more severe cases the hands become weak so that unscrewing jars or turning keys becomes difficult. If the weakness spreads to affect the knees and hips then getting out of chairs and climbing the stairs become troublesome. If the wrists, elbows and shoulders become affected then tasks such as lifting and brushing hair become a problem.

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.

Investigation of a peripheral neuropathy

The first consultation

The first essential in diagnosing the cause of a peripheral neuropathy is a careful medical history and full examination. The history needs to include medical information about all close relatives (because peripheral neuropathies may run in families), previous illnesses, alcohol consumption, diet and drugs being taken. It is a good idea to bring all your current medicines (from the doctor, chemist or health food store) to the consultation. Exposure to poisonous chemicals, especially solvents, insecticides and lead paint, is an occasional cause. Bring a list of any possibly poisonous chemicals with which you have contact to the consultation.

The consultation includes a full medical examination and careful testing of the nervous system. The consultation usually narrows down the long list of possible causes to one or two likely culprits but confirmatory tests are almost always needed. If the diagnosis does not quickly become clear a larger number of tests may be needed.

Nerve conduction tests

Most patients with a peripheral neuropathy will be referred to a consultant neurophysiologist for nerve conduction tests, often called an EMG (short for electromyogram). This test involves stimulating the nerves in the forearm and lower leg with little electric shocks. The recording electrodes are small pads on the muscles and sensory nerves in the hands and feet. The doctor (and you if

you want) can see the results on a television screen. A computer helps calculate how many nerve fibres are working and how fast they are conducting their messages. In axonal neuropathy there are too few nerve fibres and the remaining fibres conduct more or less normally. In a demyelinating neuropathy the nerve fibres do not disappear but they conduct too slowly. Sometimes it is necessary to record the electrical activity in the muscles with a very fine needle. The pattern of electrical activity can show whether the fault really lies in the peripheral nerves or somewhere else, possibly in the muscles or the spinal cord.

Urine test

This is a routine part of a thorough medical examination. It shows up diabetes and kidney disease.

Blood tests

Blood tests can diagnose lots of diseases. Here are some common ones:

	<u>Test</u>	<u>Conditions detected</u>
Haematology	Blood count	Anaemia
	Sedimentation rate	Inflammation
	Vitamin B12	Vitamin deficiency
Biochemistry	Kidney function	Kidney failure
	Liver function	Alcoholism
	Thyroid function	Thyroid deficiency
	Serum proteins	Abnormal proteins
Immunology	Autoantibodies	Autoimmune diseases
Genetics	Special DNA tests	Hereditary neuropathies

X-rays

A chest X-ray is often needed as part of a thorough medical investigation. Various sorts of inflammation in the chest can cause a peripheral neuropathy. In smokers the possibility of lung cancer may have to be considered.

Lumbar puncture

In acute neuropathies and in severe chronic neuropathies a lumbar puncture is helpful. This involves coming into hospital for the day. You have to lie on your side and the doctor gives you a local anaesthetic injection into the lower part of the back. Then he or she pushes a very fine needle through the numb area of the skin into a large hollow space in the spine. This allows collection of the spinal fluid, which bathes the nerve roots. The cell and protein content of this spinal fluid help diagnose inflammation. Most hospitals ask you to lie flat for an hour or two after but it is not usually necessary to stay in hospital. Lumbar puncture sometimes causes headache for a day or two. The headache goes away if you lie down.

Nerve biopsy

If the diagnosis has not become clear from the other tests, a nerve biopsy may be necessary. It needs a local anaesthetic and involves a cut about an inch long on the outer side of the ankle. It is best to rest in bed for a day or two afterwards, not necessarily in hospital, and to avoid strenuous exercise for at least two weeks. The stitches usually come out after 10 to 14 days. The test is only done as a last resort because it may cause pain on the side of the heel and foot for several weeks. This only happens in about 10% of cases and is less likely if the foot is very numb in any case.

Causes of a peripheral neuropathy

Many diseases can cause a peripheral neuropathy and this list shows only some of the most important.

Some important causes of a peripheral neuropathy:

- Diabetes mellitus
- vitamin B12 deficiency
- underactive thyroid
- kidney failure
- alcoholism

- Guillain-Barré syndrome
- chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
- vasculitis (inflammation of blood vessels)
- paraproteinaemia (abnormal blood protein)
- hereditary motor and sensory neuropathy (Charcot-Marie-Tooth disease)
- idiopathic axonal neuropathy.

Here are some examples:

Guillain-Barré syndrome

This is an uncommon acute neuropathy which usually affects the motor more than the sensory nerves. It reaches its worst within one or two weeks, four weeks at the most. It should be treated as an emergency. Most people make a very good recovery.

Contact the GBS Support Group.

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

This is an uncommon chronic neuropathy which also usually affects the motor more than the sensory nerves. It lasts for several months and may disappear on its own or with treatment and then come back. In about half the cases it clears up in the end.

Contact the GBS Support Group.

Vasculitis (inflammation of blood vessels)

This usually occurs as part of another disease affecting blood vessels in several parts of the body. Examples are rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosa and Churg-Strauss syndrome. Churg-Strauss syndrome is worth special mention because it commonly affects the peripheral nerves. It causes asthma and produces an acute peripheral neuropathy with the pattern of multiple mononeuropathy.

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Stuart Strange Vasculitis Trust 8 Wilsford Close, Wigton, Leicester, LW18 2RR Tel: 01662 881335 E-mail: d.smith32@ntlworld.com Web site: www.vasculitis-uk.org

Churg-Strauss Syndrome International Support Group Gary Todd, European Coordinator Lee School House, Long Framlington, Morpeth Northumberland NE65 8JG Tel: 01669

Paraproteinaemia (abnormal blood protein)

Sometimes one family of antibody-producing bone marrow cells gets out of control and churns out large amounts of exactly the same antibody. This antibody, also called an immunoglobulin, may damage the nerve fibres. This may either cause a peripheral neuropathy, a bit like CIDP, or a rather mild and very slowly progressive sensory peripheral neuropathy. Treatment is available but may not be necessary because it is so mild.

Contact the GBS Support Group.

Hereditary motor and sensory neuropathy (Charcot-Marie-Tooth disease)

It is quite common for peripheral neuropathy to run in families. Hereditary neuropathies usually show up during childhood or adolescence with difficulty running, high foot arches and toes curling. The foot problems may make it difficult to buy comfortable shoes. Often hereditary peripheral neuropathies are so mild that people do not realise they are affected. Occasionally it does cause slowly progressive weakness of the ankles and then the hands that may affect everyday activities. There are different patterns of inheritance but the commonest, hereditary and motor sensory neuropathy type 1, is inherited as an autosomal dominant condition. This means that it is passed on from parent to child. Each child, regardless whether the child is a boy or a girl, has a 50% chance of being affected.

Contact: CMT United Kingdom Mrs Karen Butcher, Secretary PO Box 5089, Christchurch, BH23 2WJ Tel: 0870 7744314 E-mail: secretary@cmt.org.uk Web site: www.cmt.org.uk

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, idiopathic peripheral 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.

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Information

Guillain-Barré Syndrome

Introduction

This guide is written by neurologists and other specialists who have a particular interest in Guillain-Barré syndrome (GBS). It is intended for patients who have been told that they have, or may have GBS, and for their relatives and friends. It is quite detailed and should be read after you have read the *Quick Guide* which gives you a rapid overview of the disease and should answer your immediate questions. (The *Quick Guide* is available on the GBSSG of the UK Web site.) It has to be honest and is meant to be reassuring. The information contained in this book is an accurate and up to date account of GBS. Situations may arise in which you receive apparently conflicting opinions and information from different doctors and health care workers about various aspects of GBS. Unfortunately the book cannot respond in words to the conflicts or concerns that this information may cause. Consequently if you do not understand or are worried by the information offered here, you must ask your medical specialist to explain. Don't be scared to quote from this book if you feel intimidated or neglected. Any good doctor should be willing to listen and to explain.

What is GBS?

GBS is an uncommon illness causing weakness and **loss** of sensation that usually recovers completely after a few weeks or months. It is named after two French physicians, Guillain (pronounced Ghee-lan) and Barré (pronounced Bar-ray), who described it in 1916 in two soldiers who were affected by a paralysis but later recovered. It affects about one person in 40,000 each year, i.e. 1,500 persons altogether each year in the United Kingdom. It can occur at any age from infancy onwards but is slightly more common in the **old**; it is more common in men than in women; it is not hereditary; it is neither passed onto children nor is it infectious and it is not caught from or transmitted to anybody else. However, it does often develop a week or two after a throat or intestinal infection.

What are the symptoms?

The first symptoms are usually either tingling (pins and needles) or loss of feeling (numbness) beginning in the toes and fingers. Legs feel heavy and wooden, arms feel limp and hands cannot grip or turn things properly. These symptoms may remain mild and clear up within a week or two without need for hospital admission but most people need to be admitted to hospital. At the earliest stage, it may be difficult for the patient to persuade the doctor that there is anything physically wrong. Within a few days it is all too obvious that something has gone wrong: legs simply will not bear weight, arms become very weak and the doctor finds that the tendon reflexes have disappeared.

How is GBS diagnosed?

The diagnosis of GBS is made from the clinical history (the story you tell your doctor) and medical examination, supported by laboratory tests. This means that the doctor will try to work out whether the history and clinical examination fit into the pattern of GBS. The doctor will particularly want to know of any recent possible infections or vaccinations, toxin exposure (such as insecticides or solvents), alcohol intake, tick bites, family history of nerve disease or symptoms of any coincidental illnesses such as diabetes (thirst, frequent urination, weight loss). Your answers to these questions might support the diagnosis of GBS or lead to a different diagnosis.

Investigations will normally include blood tests, a lumbar puncture and electromyogram (EMG). The lumbar puncture involves lying on one side and having a needle inserted under local anaesthesia between the vertebrae into the sac of cerebrospinal fluid that surrounds the nerve roots at the base of the spine. The idea is worse than the procedure really is and it does not usually hurt. In most GBS patients, the cerebrospinal fluid contains much more protein than usual while the cell content remains normal. If different changes are found, the doctor has to review the diagnosis with even more care.

The electromyogram, or EMG, is an electrical recording of muscle activity and is a very important part of making the diagnosis of GBS. It is not done in all hospitals and may therefore require the patient to be transferred to a specialist unit where the test is available. If a nerve is stimulated with a brief electrical pulse (felt like a sharp tap or jolt), muscle activity can be recorded and the speed at which the nerve conducts electricity (the nerve conduction) can be worked out. Often in GBS, nerve conduction is slowed or even blocked altogether. The test usually lasts about half an hour. Some patients find the electrical stimulation rather uncomfortable but it is entirely harmless.

What happens next?

The worst degree of weakness is usually reached within four weeks and always within six weeks. Some patients deteriorate very rapidly to a state of severe paralysis over the course of a few days but this is uncommon. The patient then enters a plateau phase that usually lasts a few days or weeks during which the course of the disease seems stationary. Most people are so weak during this stage that they are confined to a hospital bed where rest is probably a good thing. However, it is very important to keep all the joints moving through a full range to stop them stiffening up. The physiotherapist is in charge of this physical therapy and will be pleased to advise relatives and friends on what they can do to help.

Is GBS Painful?

Unfortunately, some patients get a lot of pain during GBS, particularly in the spine and in the limbs. Other patients report GBS as an entirely painless experience, even when severely paralysed. Pain may come from the inflammation of the nerves themselves, from the muscles that have temporarily lost their nerve supply, from stiff joints, or simply because the patient is lying in an uncomfortable posture and is too weak to move into a more comfortable position. To combat the pain, the doctors will prescribe painkillers and the nurses and physiotherapists will help with repositioning and physical therapy. It helps to know that some pain is common in GBS. This pain should disappear as the condition improves and the occurrence of pain does not mean that anything else is going wrong.

Do patients need intensive care?

This subject and other items concerning GBS patients in intensive care are more fully detailed in a companion booklet entitled *The GBS Patient in Intensive Care*. A brief summary is enclosed here.

Since a patient with GBS can deteriorate rapidly, it is essential to treat him or her as a medical emergency initially. Once the progression of the illness is established, the doctors will be in a better position to judge whether or not the GBS patient will need to be admitted into an intensive care unit (ICU, sometimes called an intensive therapy unit or ITU). The remainder of this section is directed only towards the patients who are transferred to an ICU.

About 25% of GBS patients have weakness of the breathing, swallowing and coughing muscles and have to be placed on a machine that will take over their breathing called a ventilator or respirator. This process is called artificial ventilation. In addition to taking over the breathing, patients undergoing artificial ventilation have a tube placed in their throats, called an endotracheal tube, which prevents fluids in the mouth and acid in the stomach from 'going down the wrong way into the lungs. If stomach acids find their way into the lungs they can cause severe damage and your doctors and other staff will do everything possible to prevent this from happening.

Admission to an ICU is less worrying than it sounds. Although occasional GBS patients may be admitted to ICU for observation only, it is normally the case that patients on ICUs are placed on an artificial ventilator to take over their breathing. Under a short general anaesthetic, the connection to the ventilator is made to a tube placed in the windpipe (trachea) via the nose or mouth. This tube, the endotracheal tube, can be left in place for a week or two. If artificial ventilation is required for longer, a surgeon may make a small opening, called a tracheostomy, into the windpipe at the base of the throat, just below the 'Adam's apple'. This is more comfortable for the patient and permits artificial ventilation for as long as necessary. The tracheostomy is also performed under a general anaesthetic. Fortunately in GBS, artificial ventilation is rarely necessary for more than a few weeks and the majority of patients do not need artificial ventilation at all.

When ventilation is no longer needed, the tracheostomy tube can be removed quite painlessly. The wound closes in a few days and eventually leaves a small scar below the line of the collar.

Intensive care in recent years has become a very sophisticated part of medicine that has enormously improved the care of severe GBS. To make this possible, pulse, blood pressure, temperature and blood chemistry have to be measured often. The pulse will be recorded by monitoring the heart beat (electrocardiogram) on a video monitor to detect abnormalities that may need treatment. Patients may need infusions into veins to provide fluids and give drugs. A tube called a catheter is placed in the bladder to drain the urine. Another tube, called a nasogastric tube, may be passed through the nose into the stomach to provide nutrition because swallowing will be impossible. Constipation can be a troublesome problem at first but eventually nurses and patients invariably work out a regime of laxatives and suppositories that works.

Communication can be a problem for a patient who is unable to talk but with winks, nods, communication cards (the Group's own cards have been distributed to all ICUs and should be available - otherwise advise us) and, above all patience it is usually possible to get the message across. If the intensive care regime seems tedious, it is worth remembering that modern intensive care has reduced the mortality rate of GBS considerably. Fortunately, death from GBS is now a rare event, occurring in around 1 in 20 cases. Death tends to occur more commonly in elderly people severely affected by GBS and with other medical illnesses such as heart, lung or kidney disease. Like any other illness, unexpected complications can arise. Death is more likely to be a result of a complication rather than GBS itself.

How long does it take to recover?

Eventually the numbness begins to recede and strength begins to come back. Once it is clear that this is a genuine improvement rather than wishful thinking, there is some cause for cautious rejoicing because improvement is likely to continue steadily. About 80% of the patients recover completely in that they are up and about walking within one year, and often much earlier than this. The time taken for recovery to occur is very variable. Sometimes it is only a week or two but most people remain affected for between three and six months.

The patients who do not recover completely may be left with minor degrees of weakness, numbness and sometimes discomfort that do not seriously interfere with their lives. A few however are left so disabled that they cannot resume their former occupations. This is usually because of residual weakness of their arms and legs so that manual work and walking are impaired. It is uncommon to be left dependent on a wheelchair for life but this unfortunately does occur in some cases. Improvement is fastest during the first few months but some patients report continued gradual improvement even after a year or two has elapsed.

What causes GBS?

The disease is due to inflammation of the peripheral nerves, often termed “neuritis”. It is like an “- it is” anywhere else in your body: an angry redness and swelling that stops the organ in question from working properly. For example, laryngitis (inflammation of the larynx) leads to the loss of voice. The peripheral nerves are like the electrical cables around your house. They connect the central nervous system (i.e. the 'mains) to the muscles and to the sense organs in the joints and skin (i.e. the “appliances”). When these cables are damaged or cut, the appliances stop working because they have no electrical power, although are in themselves undamaged. Because many nerves are inflamed, GBS is called a 'polyneuritis”. The most likely explanation for the inflammation is that immune cells called lymphocytes start attacking the nerves in error, instead of concentrating their energies on fighting off infections. This mistake in the immune system is an own goal you could do without! It is believed that the immune system has been tricked into making this mistake by an infection that often precedes GBS. Eventually the immune system realises its mistake and corrects it by either killing off the renegade lymphocytes or discharging them from the front lines of its army, thus stopping the attack on the nerves. A disease in which the immune system attacks its hoses own body is called an autoimmune disease and GBS is one of many diseases affecting the nervous system in this category.

Is there more than one type of GBS?

Yes. Perhaps it is a good idea to understand that GBS is a clinical syndrome (defined as an aggregate of symptoms) rather than a specific individual illness. In the majority of GBS cases, when the nerves become inflamed and demyelinated, the syndrome is due to 'acute inflammatory demyelinating poly [radiculo] neuropathy or AIDP. Fortunately for GBS sufferers in this AIDP category, the part of the nerve attacked is the insulating sheath around nerves fibres termed myelin, equivalent to the plastic coating around electrical cables. This myelin sheath can be replaced by the myelin-forming cells, named Schwann cells, after Dr Schwann who described them

Usually the conducting core of the nerve, equivalent to the copper core within electrical cables and called the axon, is not damaged. In the AMAN (acute motor axonal neuropathy) and AMSAN (acute motor and sensory axonal neuropathy) forms of GBS, the axons are damaged too. Although they can regrow, recovery takes longer and may be incomplete. Patients with AMAN or AMSAN may therefore make poor recoveries.

In some cases the illness may run a longer course than usual and become a chronic illness. This chronic version of the aforementioned AIDP is called CIDP (where C = chronic etc) and is described later in this booklet.

A variety of the acute condition is Miller Fisher syndrome (MFS) which is also described later. There are several other very rare conditions that are categorised as clinical variants of GBS; often they do not exhibit the full range of symptoms of the 'classic' description.

Can you tell me more about CIDP?

CIDP is less common than acute GBS (about 1:10) and most people reading this booklet need not bother with this section.

Like GBS, CIDP is an autoimmune disease of the peripheral nerves. Symptoms experienced by patients with both conditions are very similar.

CIDP is only distinguished from GBS by virtue of its pattern of progression. GBS is always defined if the low spot is reached within four weeks (and sometimes up to six weeks) although it typically happens within a few days. If the initial progressive phase lasts longer, and usually it is much longer, then the illness is called CIDP. Some CIDP patients are initially diagnosed with GBS and only when the deterioration continues over an extended period, or when one or more relapses occur after a period of improvement, is the illness reclassified as CIDP.

Although CIDP is a chronic condition, several different treatments are thought to be helpful. They all act by suppressing the damaging autoimmune response. Examples are steroids, azathioprine, 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 demyelinated nerves can be repaired, that treatment is available and that some patients get better without treatment.

Can you tell me more about Miller Fisher syndrome?

About 5% of GBS sufferers have Miller Fisher syndrome (MFS) which was described in 1956 by Dr Miller Fisher. He described patients with paralysis of the eye muscles, incoordination (ataxia) of the limbs and loss of tendon reflexes but no weakness in the arms or legs.

Strictly speaking, that and only that, is MFS. The connection with GBS comes because some GBS patients have paralysed eye muscles too. Consequently, MFS and GBS can overlap. Recently, special antibodies have been found in patients with MFS and in patients with GBS with eye paralysis but not in other GBS patients. These antibodies may be the cause of the eye muscle paralysis.

Can I get a second attack of acute GBS?

The bad news is 'yes' but the good news is that the odds are against it; a figure of 3% has been estimated. This should not be confused with the chronic condition CIDP (see previous page) but some authorities do in fact reclassify people who have a second acute attack as having CIDP even though the second attack may have occurred many years after the first.

Original text by Professor Richard Hughes.

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Axonal and “Severe” GBS

It had been known for some time that in severe cases of GBS, a 'bystander' effect of the demyelization of the nerve could be damage to the nerve core or axon. In 1986, Feasby et al [Brain 1986 Dec; 109 (Pt 6):1115-26] reported autopsy studies on a patient with a clinical diagnosis of GBS and who had died that showed severe axonal degeneration in nerve roots and distal nerves without evidence of demyelization. It was suggested that this might represent a variant of GBS characterised by an acute axonal neuropathy.

In 1995, Griffin, Ho et al reported on their findings after investigating the yearly epidemic of GBS amongst children in northern China [Brain 1995 Jun;118 (Pt 3):577-95, 597-605]. Twelve autopsied cases were studied. Three of the twelve cases showed the same characteristics of classic. Of 129 Chinese patients who were studied, 65% had the axonal form, 24% the demyelinating form and 11% could not be classified. One batch of 38 patients (55% axonal 32% demyelinating, 13% unclassified) was tested for antibodies to the bacterium *Campylobacter jejuni*. Sixty-six percent of the 38 showed evidence of recent *Campylobacter jejuni* infection compared with 16% in the control).

It did not take long for the axonal neuropathy as described by Feasby et al and the 'Chinese paralytic syndrome' to be regarded as one and the same and it was quickly recognised that *Campylobacter jejuni* was probably the most common trigger for GBS in the West as it seemed to be in China. [Hughes RA, Rees JH J Infect Dis 1997 Dec;176 Suppl 2: S92- 8]

In 1997, Ho et al reported [Neurology 1997 Mar;48(3):717-24] on the mechanisms of paralysis and recovery during AMAN. The most severe cases showed degradation of motor axons affecting the ventral roots as well as the peripheral nerves. In contrast, a patient with the characteristic findings of AMAN recovered quickly after plasmapheresis. A sural nerve biopsy proved normal but a biopsy at a neuromuscular junction showed denervation (possibly explaining the Chinese paradox). Antibodies have also been found to be binding to the nodes of Ranvier (between the myelin segments) preventing transmission. There are clearly different mechanisms at work here: one resulting in a slow and incomplete recovery and another resulting in a rapid recovery. Note: Chinese AMAN patients have been found to recover at an identical rate as Chinese AIDP patients suggesting they fell into the latter category.

So while some patients with 'axonal GBS' may recover quickly, others have considerable axonal damage. They will be joined by those who have bystander axonal damage as a result of AIDP (and indeed CIDP). The only proven treatments are plasma exchange and IVIG (AIDP) plus corticosteroids (CIDP). A problem arises because while demyelization appears to be effectively and promptly repaired by remyelination, axonal degeneration can cause severe, persistent disability.

[Hughes et al Mult Scler 1997 Apr;3(2):88-92].

Information **The Guillain-Barre Syndrome Patient in Intensive Care**

Introduction

Around a quarter of GBS sufferers are admitted to intensive care units (ICUS, sometimes called intensive therapy units or ITUS) for special care if their illness is judged severe or moderately severe. Admission to an ICU is particularly recommended for patients with weakness of their breathing, swallowing or coughing muscles. A machine called a ventilator will be introduced to take over their breathing function and to stop fluid and secretions from slipping down the trachea into the lungs where infection and lung damage may arise. It is principally for the family and friends of this category of GBS patient that this information has been produced.

It is important that you read this in conjunction with the GBS Support Group's publication, [Guillain-Barre Syndrome - a guide for patients, relatives and friends](#). This explains the illness in easily understood language. It will do much to relieve your worries. If no copy of the publication is available to you, please contact the Group by phone, fax, letter or e-mail. The contact details appear on the front cover of this booklet.

What is an intensive care unit?

This is a special unit within hospitals, staffed by medical support personnel who are specially trained in the high levels of care required by each patient.

There is nothing sinister or depressing about these units. On the contrary, they are busy and cheerful places where patients are under constant watch, day and night, and everything is done to ensure that they receive the highest level of care possible. At first sight, there appears to be a daunting amount of equipment at the bedside, but you will be surprised how soon you and your affected relative or friend come to understand the function of each piece of machinery.

Why has the GBS patient been admitted to the ICU?

The patient's heart will be monitored on a screen to watch for any irregularities. A thin tube (catheter) may be used to drain urine from the bladder.

In order to feed the patient whose swallowing ability is impaired by the GBS or made impossible by the plastic breathing tube, a special tube called a nasogastric tube will be passed through the nose and down the throat and esophagus into the stomach so that liquid food may be taken in.

Pulse, blood pressure, temperature and other vital signs will be regularly monitored.

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Airways and lungs will be kept clear by a method of suctioning, as and when required. This is an essential procedure which, when completed, gives the patient considerable relief. However, it is noisy and if visitors find this distressing or unpleasant, they should quietly leave the unit until it is over.

This all sounds a bit frightening, but remember that these procedures are all regularly used in ICUs and are essential for the patient's well being. Each support mechanism will be discarded as the GBS sufferer improves.

How does the ventilated GBS patient feel?

At first, patients are very alarmed at the new situation and surroundings in which they find themselves. However, they soon become familiar with what is going on and begin to understand the routine. A simple but careful explanation is essential to put the patient's mind at rest.

The first thing to note is that the GBS patient cannot speak and may also have a reduced or absent sense of taste and smell. Some patients will also experience visual disturbance. Hearing is rarely impeded, so the patient can generally understand and acknowledge all that is going on. However, appreciation of the surroundings may also be dampened by sedative or pain killing drugs which are often used to make GBS patients more comfortable.

Some patients do experience an increase in skin sensitivity so although touch is important, care must be taken. In some rare cases even a light touch may cause very severe pain which the patient cannot easily communicate to you.

GBS is a paralyzing illness. The paralysis is temporary but can be quite extensive and the patient is fully aware of the lack of movement. This can be both perplexing and hard to accept.

Many GBS patients are alert and acutely aware of what is going on. They feel vulnerable, isolated and locked-up inside their illness. Considerable frustration occurs because they are unable to talk whilst on the ventilator, and you may encounter some irrational or uncharacteristic behaviour. It is never easy for them to come to terms with what has happened, so do not be surprised if they are variously tearful, bad tempered or panicky.

Everyone coming into contact with the ventilated GBS patient should remember at all times that the patient is quite aware of his or her reliance upon the machinery to which they are attached. Remember too that from a mental and emotional standpoint, loss of movement and inability to speak makes patients feel fragile and vulnerable. A less than caring action or unsympathetic attitude can set the alarm bells ringing inside the silent patient.

What can I do to help?

You can do a great deal! Your first task is to understand, at least in outline, what this illness means. Speak to the doctor in charge of the case as soon as you can to get yourself into the picture. Some doctors are better than others at explanations. Don't hesitate to ask questions. Have you read the guide published by the GBS Support Group? Does the hospital know that there is a national Support Group?

Secondly, familiarize yourself as soon as you can with the ICU. Get to know the regular nursing staff who will give you a daily update on progress.

If a patient is to receive a new treatment or procedure, make sure he or she knows about this in advance and understands why it is being undertaken.

A physiotherapist may begin passive movement of the limbs whilst the patient is bed-bound. Get to know the physio and keep yourself updated on procedure and progress. There is a lot he or she can tell you

The patient cannot talk but is anxious to communicate. Make sure the speech therapist is involved in advising on communication aids. If good facial strength has been retained then lip reading will be effective. Some patients retain finger movement and can write letters in the air or on the palm of the hand. A common method of communication with a patient whose movements are restricted to the eyelids, is to use a question and answer technique with the patient answering with one blink for "yes" and two for "no", sometimes running through the alphabet until the correct letter is found. This can be improved upon by pointing to the letters on an alphabet board and asking 'is it on this line?' or 'is this the letter?' The patient can respond as before by using the eyelids or perhaps by banging his or her teeth together with the jaw muscle.

If the patient is strong enough, he or she may be able to point at an alphabet board with a finger or pointer attached with a headband. A very useful method of communication is by the use of communication cards which pre-empt many questions or comments the ventilated patient is likely to want to ask.

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If the patient is strong enough, he or she may be able to point at an alphabet board with a finger or pointer attached with a headband. A very useful method of communication is by the use of communication cards which pre-empt many questions or comments the ventilated patient is likely to make. ICUs should possess a copy of the Group's own cards. If they are not to be found, copies are available from the Group.

The GBS patient puts a lot of effort in trying to communicate and you soon find a method that works and you will become quite expert. Encourage others to understand too.

The GBS patient is socially isolated and needs to be stimulated. Make sure he or she knows the day of the week and the date. Encourage friends and family to send cards and write letters about what they are up to. If the patient cannot see TV, relate what is going on in the outside world. Read ex-tracts from a national or local newspaper. Would the patient like a cassette storybook played? Always include the patient in bedside conversation.

Financial worries may be bothering the patient, especially if he or she is a breadwinner. Get in touch with the Social Worker at the hospital who will advise on State benefits and claims. Alternatively, your local Citizens Advice Bureau dispenses free and expert advice on benefits.

Early action is essential, as many benefits cannot be claimed retrospectively. Inform the patient's employers about GBS and confirm the situation on job security. Patients worry a good deal about such matters.

As the patient's breathing improves, he or she will be gradually taken off the ventilator, starting with just a few minutes and building up from there. Patients can get quite panicky at the beginning of this procedure as they have become reliant on the ventilator and do not believe, initially, that they can breathe again without it. Reassure them that their natural ability to breathe is returning and that this is the start of getting well.

In summary

It is impossible to cover every single aspect of the GBS patient in the ICU. This is a very personal illness and each patient has his or her particular set of problems and worries to cope with. Your role is to offer love, comfort and reassurance during this difficult period. To do this effectively, you must remain calm and resolute and give constant encouragement on progress. Patients easily lose sight of how they are doing so keep yourself well informed by the medical staff. Writing a diary of daily events will help you keep a perspective on progress. You can relate this to the patient who may not realise how he or she is getting on.

Some days are better than others for GBS patients and it is hard to be a hero every day, but you must keep up a constant flow of encouragement.

For the close family, this period of the illness is quite stressful, so don't forget to look after yourself and stay well.

Original text by Sandra Stellman.

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Information

Childhood Guillain-Barré Syndrome

Introduction

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

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

*Radiculo' is sometimes omitted

Definition

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

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

Clinical course

GBS usually occurs around two weeks after a respiratory (e.g. a cold or 'flu) or a gastro-intestinal (e.g. diarrhoea or vomiting) infection. More rarely it may follow other infections such as chicken pox. We do not know why it affects a particular person as more often than not the whole family has had the infection but usually only one person will go on to get GBS. Furthermore, the person affected will not necessarily have been the most unwell with the initial illness.

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

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

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

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

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

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

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

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

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

Differential diagnosis - what else could it be?

Not all children present with the classical history described above. It is very important for everyone to feel confident with the diagnosis of GBS as this would affect the management of your child. Some children can have many signs in common with GBS but not have the condition. Thus the doctor may need to perform a number of other tests to ensure nothing is missed, especially if it may alter the way your child is treated. The investigations are likely to include blood tests, urine samples and some body swabs. It may be necessary to perform a lumbar puncture in the first week of presenting if there is any fear of ongoing infection. Rarely, a 'follow up' lumbar puncture may be needed a week later. Some children have neuroimaging of their head and/or spine (by a CT or MRI scan) when the diagnosis is in doubt. The doctor will try to keep the tests to a minimum but it is important nothing is missed. Make sure you understand what each test is for, otherwise it may be a big surprise to find your child's treatment suddenly changing.

Management

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

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

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

During the illness, it is important to keep any monitoring device the doctors and nurses advise on your child. He or she may become irritated with it, but it must remain. Most children will need monitoring of their heart rhythm (pulse), breathing (oxygen saturation) and blood pressure at regular intervals. The doctor will also be concerned about the breathing and will ask your child to blow into a machine to measure the 'vital capacity' at regular intervals. If your child is too young or weak to do this then the doctors and nurses will watch the breathing pattern carefully to check it does not become too laboured and the doctor may need to do blood tests to monitor it.

Treatment

Steroids

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

Immunoglobulins

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

Plasma exchange

Plasma exchange has also been shown to speed up recovery, but it is more complex to administer than IVIG. The child requires several large intravenous lines for this procedure and about 250mls (1/2pint) of blood are removed at a time in a closed circuit and 'washed' before re-entering the child. By doing this, an attempt is made to remove the circulating antibodies in the blood which are attacking the nerves. This would only be performed in an intensive care unit and although it is quite an invasive treatment, it is frequently performed in the above setting for other conditions.

The exchange may need to be performed on several occasions until evidence of improvement is seen.

Pediatric intensive care

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

Things you can do to help

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

Outcome/rehabilitation

The majority of children with GBS make a full recovery without any signs of having had the condition. However a small number may have some persisting problems. The commonest complaint is weakness of the hand and foot muscles. Most recovery is seen in the first few months. However children can continue to improve for up to two years after the illness. The duration of the plateau period (i.e. the time spent at maximum weakness, before recovery begins)

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

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

Recurrence

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

Variations

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

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

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

Who's who?

The team involved in your child's care are:

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

References

- Brett E M. Paediatric Neurology. 3rd edition. Churchill Livingstone. Neuromuscular Disorders: H. Peripheral Neuropathy. 1996: 4
- Ouvrier R, McLeod J G and Pollard J. Peripheral neuropathy in childhood. Raven Press. Acute Inflammatory Demyelinating Polyradiculopathy. 1990: 5; 39-49
- Korinthenberg R and Monting J S. Natural history and treatment effects in Guillain-Barré Syndrome: a multicentre study, Archives of Disease in Childhood. 1996: 74 (4);281-7

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Information Miller Fisher Syndrome

Also known as:

- Miller Fisher variant
- Fisher('s) syndrome
- acute idiopathic ophthalmologic neuropathy

In 1956, C. Miller Fisher, a US doctor, described three patients with acute external ophthalmoplegia (eye paralysis), sluggish pupil reflexes, ataxia (lack of balance) and areflexia (absent tendon reflexes). Two patients had no weakness; the other had a facial palsy and possible weakness. All three recovered spontaneously.

Because some patients with GBS had ophthalmoplegia and there were other similarities, Dr Fisher concluded that these patients had suffered a disorder akin to GBS. [Fisher CM. Syndrome of ophthalmoplegia, ataxia and areflexia. N Engl J Med 1956;255:57-65]

Pure Miller Fisher syndrome (without generalised weakness) is rare. Electrodiagnostic abnormalities found in all patients are characteristic of an axonal neuropathy or a neuronopathy with predominant sensory nerve changes in the limbs and motor damage in the cranial nerves. [Fross RD, Daube JR- Neuropathy in the Miller Fisher syndrome: clinical and electrophysiologic findings. Neurology 1987 Sep;37(9):1493-1498]

Patients described as having Miller Fisher syndrome often have a neuropathy that overlaps with GBS and demonstrate generalised weakness, sometimes paralysis, as additional symptoms.

It was sometimes proposed the Miller Fisher syndrome was caused by brainstem encephalitis. It is true that the syndrome can be mimicked by a brainstem lesion, but typical cases of Miller Fisher syndrome rarely show any evidence of brainstem abnormalities either radiologically or during post- mortem examination. When clinical or radiological brainstem abnormalities are found, the condition may be referred to as Bickerstaff's syndrome or Bickerstaff's brainstem encephalopathy.

It is possible that the tremor and ataxia symptoms of Miller Fisher syndrome are explained by a disparity between different sensory inputs and that all the clinical features are due to abnormalities of peripheral nerve function. [Ropper AH, Shahani B. Proposed mechanism of ataxia in Fisher's syndrome. Arch Neurol 1983 Sep;40(9):537-538]

Research in recent years has concentrated in identifying the antibodies that are thought to be responsible for GBS etc. It has been discovered that the antibody found in sufferers of pure Miller Fisher syndrome is also present in those patients who have the Miller Fisher/GBS overlapped condition. This particular antibody is not found in other GBS patients so it is thought that it is responsible for the ophthalmoplegia.

Although not clinically proven, treatment of Miller Fisher syndrome is much the same as 'classic' GBS though the different symptoms require modified management with emphasis on the eyes. Intravenous immunoglobulin or plasma exchange treatment is likely in all but the mildest cases. The chances of recovery are very good.

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CIDP (Chronic Inflammatory Demyelinating Polyradiculoneuropathy)

Introduction

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

*'Radiculo' is sometimes omitted

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

What is CIDP?

CIDP is defined thus:

- 'chronic' refers to the gradual course of the illness,
- 'inflammatory' means there is strong evidence that it is inflammation that causes the nerve damage;
- 'demyelinating' means that the damage is primarily to the insulating myelin sheaths around the nerve fibres; and
- 'poly[radiculo]neuropathy; 'poly' means many, ['radiculo' means root], neuro' means nerve 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 very rare disease of the peripheral nervous system involving gradual development of weakness and loss of sensation predominantly in the arms and legs.

The incidence and prevalence of CIDP are very difficult to determine because of its rarity. Various estimates put the incidence at between 75 and 250 people per year in the UK. The disease may start at any age, but is slightly more common in young adults. It is more common in men than women. For women, relapses are slightly more likely to occur during a pregnancy year.

It is not hereditary; i.e. it is not passed on to children. It is not infectious; i.e. it is not caught from, or transmitted to, anybody else. It is not a psychiatric or 'nervous' disorder.

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

Symptoms

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

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

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

Diagnosis

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

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

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

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

Essential criteria for a positive diagnosis of CIDP are:

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

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

The lumbar puncture involves lying on one side and having a needle inserted under local anaesthesia between the vertebrae into the sac of cerebrospinal fluid which surrounds the nerve roots. The idea is worse than the procedure really is and it does not usually hurt. The cerebrospinal fluid often contains much more protein than usual while the cell content remains normal if different changes are found the doctor has to review the diagnosis with even more care.

The EMG is an electrical recording of the muscle activity. If a nerve is stimulated with a brief electrical pulse (felt like a sharp tap or jolt) muscle activity can be recorded and the speed of nerve conduction worked out. Usually in CIDP nerve conduction is markedly slowed or even blocked. The test lasts about half an hour. It is only slightly uncomfortable and quite harmless.

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

Sometimes a nerve biopsy may also be performed. This involves a small piece of nerve being removed, usually from the side of the heel of the foot, to be examined in the laboratory. This allows the doctor to see any inflammation and the type of nerve damage. Having the biopsy is not painful because local anaesthetic is used, but the skin below may become sore for a week

or two afterwards. The patient may be left with some loss of sensation in a very small area on the side of the foot.

Progression

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

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

Clearly the cutoff points used are somewhat arbitrary.

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

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

What is going on?

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

The myelin helps the conduction of messages along the nerves as well as insulating and protecting the nerve.

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

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

Fortunately the myelin sheath can be replaced within a few weeks or months by the myelin-forming cells, named Schwann cells. If the nerve axons are damaged these can also regrow, but this is much slower. Research is continuing into the underlying causes and mechanisms of the disease.

Treatment

Treatment of CIDP is usually very effective with about 80% of new cases making a dramatic response to therapy, although there is no one shot curative treatment in the way that antibiotics might cure an infection. Drug treatments are generally thought to work by suppressing the autoimmune response. This in turn reduces the disabling symptoms of the disease. Examples are steroids, immunosuppressive drugs, plasma exchange and intravenous immunoglobulin.

Obviously suppressing the immune response cannot be undertaken lightly because it runs the risk of suppressing normal immune responses to infections. The decision whether to try these treatments has to be tailored by the doctor to the individual needs of each patient. However it is reassuring to know that treatments are available, that demyelinated nerves can repair themselves, and that some patients get better without treatment.

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

Steroids

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

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

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

immunosuppressive drugs

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

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

Plasma exchange

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

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

Immunoglobulin

There is increasing evidence of the effectiveness against CIDP of intravenous infusions of immunoglobulin (also called gamma globulin or antibodies). Antibodies usually react with and neutralise germs which get into the body. These are 'good' antibodies. Sometimes antibodies attack the body itself and these 'bad antibodies, or autoantibodies, may cause CIDP. However there are also anti-autoantibodies, which block these bad antibodies. It may be these anti-autoantibodies in immunoglobulin which help.

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

It is given by infusion into a vein, usually every day for five days. Each infusion takes about five hours. The immunoglobulin used in the UK has an excellent safety record. Abroad there have been very rare cases of transmission of hepatitis but considerable care is now taken in the purification and removal of any viral particles which reduces this risk to the absolute minimum

With any blood product there is always a slight risk of transmission of a new infection such as Creutzfeld Jakob disease (CJD) which has received a great deal of recent publicity. For this reason immunoglobulin from British Donors is not currently being used as a source for the manufacture of immunoglobulin until there is confirmation that there is no risk associated with it. This extra safeguard should reduce the concerns of anyone receiving this very effective treatment. There is a rare (about 1 in 40,000) risk of serious allergic reaction at the start of each infusion, so careful monitoring is essential. Some patients only need one course. Others need repeated courses.

Physiotherapy

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

The aims of physiotherapy are to:

- maximise muscle strength and minimise muscle wastage by exercise using strengthening techniques;
- minimise the development of contractures (or stiffness) around joints; a physiotherapist can advise on passive stretching techniques to help maintain full range movement at joints;
- facilitate mobility and function; sometimes, if muscles are very weak, function can be improved by the use of splints and
- provide a physical assessment which may help in planning future management

Living with CIDP

Coping with Uncertainty

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

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

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

You and your family and friends

A diagnosis such as CIDP, a chronic condition with an uncertain prognosis, may 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 musingly mature and a source of strength. Trying to keep your problems to yourself will not spare them any anxiety.

You and your doctor

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

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

Attitude to life

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

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

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

What you can do to help yourself

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

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

Regular exercise is important for overall health and should be taken according to individual limits and capabilities. Over exertion causes fatigue. However a little regular exercise will help to minimise muscle wastage and give you a good feeling of 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.

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