GBS New Zealand

GUILLAIN BARRÉ SYNDROME

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Peripheral Nerve Disorders

GBS Support Group New Zealand Trust

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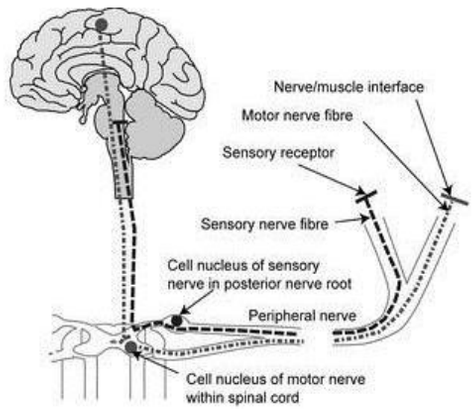
Introduction How peripheral nerves work

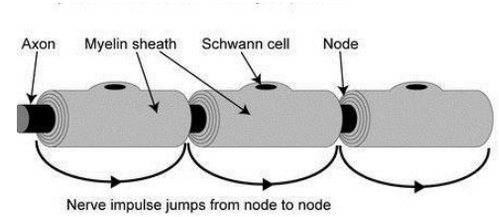
Peripheral Nerve Disorders

The peripheral nerves connect the central nervous system (the brain and the spinal cord) to the periphery (the sensory receptors and muscles). An illness of the peripheral nerves is called a ‘peripheral neuropathy’. Peripheral nerves are made of bundles of nerve fibres, which can be regarded as living telephone wires. They are kept alive by their cell bodies. The cell bodies of the motor nerves lie in the spinal cord within the spinal column or around the base of the brain. The cell bodies of the sensory nerves are in bunches, called ganglia, connected to the nerve roots on the back of the spinal cord or brain stem.



The motor nerve cell body has a long fibre called an axon, which extends from the central nervous system to the muscles. The longest axons can be as much as a metre in length, for example the nerves to the muscles in the feet. The connection between the motor axon and the muscle fibre is a

specialised nerve ending, which contains tiny packets of a chemical. The motor nerve impulse stimulates the motor nerve ending to release the chemical and make the muscle fibre contract. If a peripheral neuropathy affects the motor nerves, the muscles become weak because they do not receive the messages to move.



The cell body of a sensory nerve has two axons. One goes into the spinal cord and delivers messages to the brain. The other comes from specialised receptors in the skin, joints and muscles and transmits messages from them. The receptors sense changes in pressure, position, 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 are called unmyelinated fibres and conduct nerve impulses more slowly. They signal pain and temperature and are important for the control of blood circulation and sweating.

Different types of peripheral neuropathy

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

A peripheral neuropathy often affects all the nerves more or less together. Because the longest nerves are the most vulnerable, the feet and then the hands are most affected. Such a symmetrical pattern, affecting the feet and hands more than the hips and shoulders, is called a distal (which means farthest from the body), **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 (GBS) and in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP).

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

Nerve fibres may be damaged in several 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.

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.

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.

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, Charcot-Marie-Tooth disease 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.

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

Compiled by Richard Hughes, Siân Bensa, Rob Hadden and Andy Leitch

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
- alcohol overuse
- underactive thyroid
- kidney failure
- vitamin B12 deficiency
- Guillain-Barré syndrome (GBS)
- Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
  vasculitis (inflammation of blood vessels)
- paraproteinaemia (abnormal blood protein)
- hereditary motor and sensory neuropathy (Charcot-Marie-Tooth disease)
- chronic 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. The worst degree of weakness is usually reached within 2 weeks and always within 4 weeks. It should be treated as an emergency. Most people make a very good recovery.

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.

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 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. 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 doing buttons, 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 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:

	Test	Conditions detected
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.