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Better Outcomes

# Understanding Motor Neurone Disease

*THIS FACT SHEET explains the different forms of motor neurone disease (MND), possible causes and who it affects. It describes symptoms, diagnosis, the effects, treatments and offers care-giving suggestions.*

## What is Motor Neurone Disease?

Motor neurone disease (MND) is the name given to a group of diseases in which the muscles that enable us to move, speak, breathe and swallow fail to work normally.

## What Causes MND?

The cause of MND is unknown but researchers' theories about its origins include discussion of viruses, environmental toxins and chemicals as well as immune factors. They also discuss transmitter chemicals that control communication between nerve cells (neurones), and the way in which the neurones that control movement grow, repair and age.

**Motor system:** Muscles, and the nerves that supply them, are called the motor system. It looks after action and movement. The system fails when the neurones that control movement (motor neurones) become damaged and stop working. Without nerve impulses (messengers travelling to and from the brain and the body) to activate them, the muscles gradually waste away.

There are two different types of motor neurones. MND can affect both.

**Upper motor neurones:** Upper motor neurones are in the brain. Damage to these causes "spastic" or stiff paralysis of the muscles that they serve.

**Lower motor neurones:** Lower motor neurones are in the spinal cord. Damage to them causes floppy paralysis.

## Who Develops MND?

Each year, about one in 50,000 Australians develops MND. MND can affect anyone but is slightly more common in men than in women and among people in their 50s and 60s. MND is not contagious or infectious.

**Hereditary MND:** Most cases of MND are not inherited but in some cases there is a genetic link with at least one other case of MND in the family. In 1993, researchers identified mutations (changes) in a gene called SOD1, on chromosome 21. These mutations are believed to be responsible for about 20 per cent or one in five of these genetically linked cases.

It is possible for people at risk to receive genetic testing for these particular gene mutations but only after talking with a genetic counsellor. Testing cannot rule out the presence of other mutations, as yet unknown, that may also cause MND.

## 4 Main Types of MND

- > Amyotrophic lateral sclerosis (ALS)
- > Progressive muscular atrophy (PMA)
- > Progressive bulbar palsy (PBP)
- > Primary lateral sclerosis (PLS)

**ALS:** ALS is the most common form. It affects upper and lower motor neurones. The limbs stop working properly and there is muscle weakness and stiffness, over-active reflexes and emotions may change rapidly.

**PMA:** PMA affects lower motor neurones and is characterised by muscle wasting, weakness and twitching and loss of weight.

**PBP:** PBP affects the muscles used for speech and swallowing. Nerves that control these muscles are in the “bulb” or lower part of the brain. “Palsy” means paralysis.

**PLS:** PLS is rare. Upper motor neurone damage causes stiffness and limb paralysis.

## Symptoms

Early symptoms of MND are mild and the pattern of symptoms varies from person to person.

Usually the condition begins by affecting only one part of the body before gradually spreading. In others, symptoms are widespread from the start.

Symptoms include:

- > Cramps
- > Slurred speech
- > Swallowing difficulties
- > Slowed movement
- > Muscle stiffness, twitching or jerking
- > Difficulty walking or holding objects
- > Emotional responses trigger easily

## Diagnosis

It can be difficult to diagnose MND because it resembles several other conditions. Doctors may need to keep the person under review for weeks, months or years to be sure. The person will probably be referred to a neurologist – a specialist in conditions that affect the brain and nervous system.

The doctor may order various tests, including some to rule out other conditions. These may include nerve conduction studies (NCS) and electromyography (EMG), which look at how nerves are functioning by stimulating them electrically and recording the muscle activity that results. The EMG also involves putting a fine needle into the muscles.

## Treatment

At present there is no cure for MND, although research continues throughout the world and there has been encouraging progress. In Australia a medication has been approved for the treatment of amyotrophic lateral sclerosis (ALS) – the most common form of MND. This drug is Rilutek (riluzole), which is available at a subsidised price on the Pharmaceutical Benefits Scheme. There are some strict criteria governing which people can receive subsidised Rilutek, and it is best to discuss these with a neurologist.

Researchers are developing and trialling drugs to slow down the progression of the condition.

Your neurologist or the Motor Neurone Disease Association can give you the latest information.

## How MND Progresses

In most cases, MND does not affect intellect, memory or the senses (sight, hearing, taste, smell, touch), although it occasionally causes dementia.

Bowel and bladder are not usually affected, but constipation can be a problem when people become less mobile or have to change their diet because of swallowing difficulties.

Most people will eventually have difficulty moving about and many will have difficulty with speech and swallowing.

## Caring for Someone with MND

A great deal can be done to help someone with MND maintain their quality of life. An alert rehabilitation team plays a vital role by stepping in to prevent problems before they occur and supports you in caring for your family member at home for as long as possible.

From the early stages, therapists can suggest ways to avoid complications and advise on aids, equipment and services to make life easier.

People with MND tire easily. It's important to learn how to conserve energy, simplify work, establish a regular sleeping pattern, plan activities in advance and to allow for regular rest periods. Occupational therapists can help in this area.

The speech pathologist on the team will help you to manage speech and swallowing difficulties. The team may also provide counselling for the person and the family, and can help organise respite care.

## Managing Pain

**Massage:** Some people with MND may find their skin becomes unpleasantly sensitive. Others experience cramps. As the disease progresses, limbs become less mobile and require extra care. Try gentle massage on the affected areas and keep them warm until the pain subsides. Gently moving the affected limb can relieve stiffness and joint pain.

**Joint problems:** Muscle weakness can put strain on a joint, causing pain and stiffness. Inactive muscles in the shoulder area may also result in a "frozen shoulder", which causes painful movement. Never pull on the person's arms when helping them to move and don't leave a weak arm hanging.

**Postural issues:** The loss of normal posture control may cause pain in the muscles of the neck, shoulders, hips or knees. Carers need to know how and when to readjust the person's position to minimise this problem – your physiotherapist or community nurse can help. Electrically operated beds and chairs are useful and special cushions and mattresses provide extra padding, particularly when someone has lost weight.

**Applying heat:** Warmth can help relieve pain. Try hot packs or even a warm bath. TENS therapy – Transcutaneous Electric Nerve Stimulation – may also help. Talk to your physiotherapist about this. TENS machines can be hired or obtained on loan.

**Relaxation:** Relaxation techniques help to relieve anxiety and have been of great value to many people with MND, especially those with breathing or swallowing problems. Try the self-help section of a bookstore or call the Motor Neurone Disease Association for more resources. Doctors can also prescribe a range of medications for pain and anxiety.

**Acupuncture:** Some people find that acupuncture helps to relieve pain. A suitably qualified acupuncturist will be registered either with the Australian Medical Acupuncture Society or with one of the peak organisations in the field of Chinese Medicine, such as the Australian Acupuncture and Chinese Medicine Association, the Federation of Chinese Medicine Associations or the Register of Acupuncture and Traditional Chinese Medicine Incorporated.

Make sure the acupuncturist uses sterilised needles and keep your doctor informed about all your treatments.

## The Outlook for MND

MND affects each person differently: the rate the condition progresses, the muscles it affects and the length of time people survive MND varies greatly. While the symptoms never actually improve, some people remain stable with relatively little disability for many months or years.

In the most severe cases, a person might live for only a few months after diagnosis. Others live two or ten years or more. In rare cases, the condition seems to stabilise and progress no further.

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