MND Factsheet 1 What is Motor Neurone Disease?

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Introduction

"Motor Neurone" is the name given to the type of nerve that carries messages from the brain to the muscles, telling the muscles when and how to work.

Motor Neurone Disease is the name given to a related group of diseases affecting the motor neurones of the nervous system, commonly referred to as MND (or ALS), which are degenerative, progressive and incurable.

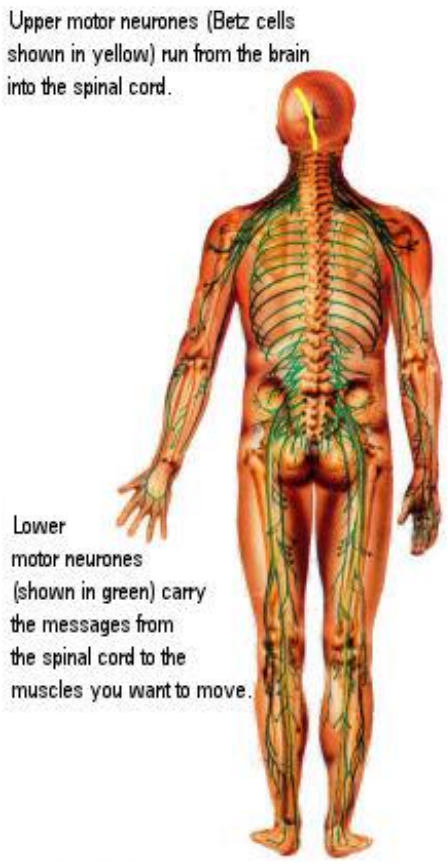
The disease can present in different ways depending on the location of the neurones involved. The cause of MND is not known, but a great deal of research is being carried out and advances are being made in understanding the disease process and the way in which motor neurones function.

Types of Motor Neurone

Within the brain there are specialised areas which deal with particular functions; the area of the brain concerned with the conscious control of voluntary movement is called the "motor area." This area contains large neurones, called "upper motor neurones", which send connections, called axons, to the spinal cord. Another type of motor neurone (called lower motor neurones) grows out from the spinal cord to connect to the muscles. When you decide to make a movement the message is carried from the brain to the spinal cord by the upper motor neurones. The lower motor neurones, in turn, carry the message from the spinal cord to the muscle causing the muscle to react.

Doctors differentiate between upper and lower motor neurones since they have slightly different functions in the body and some diseases can be specific about which types of motor neurone they affect. The Polio virus, for example, specifically affects only lower motor neurones.

If lower motor neurones are progressively damaged then the response of the muscle they control will become weaker. With time, as the number of damaged connections increases, a threshold is reached where the muscle no longer responds.



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When upper motor neurones are progressively damaged, but there is no damage to the lower motor neurones, the messages are unable to leave the brain so losing control of the affected muscle,

MND
MND damages and destroys the neurones that carry messages from the brain to the muscles. Once these motor neurones have been damaged messages cannot reach the muscles leading to weakness and, eventually, paralysis.

Approximately 200 people are diagnosed with the disease in Scotland each year.
For a small number of cases (about 5-10%) the condition runs in the family. (See Factsheet Number 2 “The Genetics of MND” for more information on this.) However, in the vast majority of cases there is no known family history.

It must be stressed that MND affects people in different ways. Not everyone will suffer the same symptoms and the rate of progression of the disease will differ between individuals while the senses will usually remain intact.

MND can manifest in different ways depending on the location of the predominant motor neurone damage.

Amyotrophic lateral sclerosis (ALS) results from damage to both upper and lower motor neurones and is the most common form affecting about 65% of people with MND. ALS is characterised by both muscle weakness and stiffness, over-active and under-active reflexes and rapidly changing emotions; the limbs may cease to work properly.

Progressive Bulbar palsy is the initial diagnosis in about 25% of MND cases and is caused when the motor neurones that control speech and swallowing are affected. The nerves that control these functions are located in the base of the brain.

Progressive muscular atrophy (PMA) is caused by damage to the lower motor neurones only and is characterised by muscle wasting (atrophy) and weakness, loss of weight and muscle-twitching (fasciculations.) 7.5% of people with MND receive this diagnosis.

Primary lateral sclerosis is a rare form of the disease affecting about 1% of people with MND and results from damage to the upper motor neurones only. Damage to these neurones results in increased muscle tension causing stiffness and spastic paralysis of the limbs.

Diagnosis
MND can be difficult to diagnose. There is no single specific test for MND. Investigations are usually carried out to eliminate other possible conditions and help confirm the clinical diagnosis.

Your Neurologist will probably arrange for a number of tests such as Electromyogram (EMG) and Nerve Conduction studies (NCS), which test the muscle and their nerve connections. Often blood tests, and scans, such as Magnetic Resonance Imaging (MRI), may also be used to exclude other possibilities and support the diagnosis.

How does the disease progress?
The symptoms and signs may be very slight in the early stages of the disease.

There could be spontaneous twitching of the weakened muscles (fasciculations), general fatigue, or loss of weight. Other early symptoms could include cramp in the affected muscles. For some there may be slight difficulty in speech or swallowing or shortness of breath if the breathing muscles are weakened.

Since MND is a progressive disease, muscle weakness increases with time, leading to increasing disability. Joint stiffness may also cause discomfort.

Muscles weakened by MND do not recover, but there may be periods of weeks or months where the illness does not appear to progress.

Some people with MND may have a tendency to cry or laugh rather easily. This condition, which is called emotional lability, can be distressing. It is becoming recognised that MND is sometimes accompanied by behavioural and cognitive changes.

What is not affected?

It is important to emphasise that the person with MND is generally in control of his or her intellectual faculties, feelings and emotions. MND does not affect the senses (touch, taste, sight, smell and hearing) and the sense of feeling in the affected parts remains normal. Bladder and bowel muscle function will remain normal, so incontinence is not normally a feature of MND However, drugs prescribed to control symptoms or loss of the ability to "push" with the tummy muscles may disturb bowel and bladder function as the illness progresses making going to the toilet more difficult than usual. Sexual desire and function are not directly affected.

How are MND Symptoms Treated?

Many of the symptoms can be helped with the proper combination of medical treatment, specialised equipment, nursing and psychological support.

Riluzole is the only drug approved for the treatment of MND where, for most people, it slows the progression of the disease delaying the onset of more serious symptoms and prolonging life by a matter of months.

Medicines can be given to help relieve symptoms like cramps, pain, stiffness, and excessive salivation. Constipation can be helped with laxatives and diet change. Practical equipment is available to assist with weak joints, poor cough, mobility problems and communication difficulties. As the disease advances, some people with MND may lose the ability to communicate easily. (See factsheet 30, *Communication Strategies* for more information.)

Help is Always Available

To ensure people with MND and their carers receive the best care possible NHS Scotland funds specialist clinical care advisors to provide support for people with MND. They are called The Scottish MND Clinical Specialist Team, and they are based in hospital neurology departments across Scotland.

People with MND can arrange for the MND Clinical Specialist to visit them, their carers and their family at home. The visit allows time for everyone to discuss concerns and any outstanding questions to be answered.

The MND Clinical Specialists are an excellent source of practical advice; they can help access services provided by the NHS and Local Authority Social Work Departments ensuring that any necessary equipment and support is in place as soon as it is needed.

The MND Clinical Specialist may also know of easy solutions to minor problems you consider aren't worth mentioning. Remember; your MND Clinical Specialist can be approached at any time, even if some time has elapsed from the initial diagnosis of MND or their first visit.

Further Information

Factsheet 22	Riluzole
Factsheet 23	Tools to Help You

Factsheet 30 Communication Strategies
Factsheet 31 Sources of Support

Introductory books available to borrow from MND Scotland's library

- Motor Neurone Disease; A Family Affair by Dr David Oliver, Sheldon Press 2011

- Navigating Life with Amyotrophic Lateral Sclerosis by Drs Mark Bromberg and Diane Banks Bromberg
  Oxford University Press and American Academy of Neurology 2017