

Motor neurone disease

Motor neurone disease (MND) is a rare condition, which is caused by the breakdown of the nerve cells in the brain that control the muscles. Unfortunately, there is currently no cure, and most people with MND die from it within a few years. However, research is underway to understand the causes, and develop a cure.

MND usually begins between the ages of 50 and 70, and only around two people in 100,000 in the UK develop the disease.

It affects the muscles used to move (voluntary muscles), but not the nerves dealing with sensation, so there is no numbness or pins and needles. The parts of the brain dealing with intelligence and awareness also remain unaffected.

There are three main types of MND, which affect different nerves:

- amyotrophic lateral sclerosis (ALS), which affects about half of people with MND,
- progressive muscular atrophy (PMA),
- bulbar palsy.

The cause of MND isn't known. Some

symptoms are like polio, so it was originally thought to be due to a virus. However, no virus has ever been discovered, and there's no evidence that it can spread from person to person.

Some types of MND run in families, so a genetic link has been suspected, but even the most common genetic forms are incredibly rare, occurring in only one in every 250,000 people.

In these genetically-linked cases, MND is known as an autosomal recessive condition, which means that both parents have to be carriers of the faulty gene to pass it on to an affected child.

Symptoms

MND usually begins very gradually, and there may just be a feeling of tiredness

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to start with. Clumsy fingers and a weak grip are often the first sign of the muscle problems. After a while, turning door handles becomes difficult. This is usually followed by difficulties in speech and swallowing.

When the foot muscles are affected, raising the foot with each step can be difficult, causing the feet to drag on the floor ("foot-drop"). The muscles of the chest wall may be affected, leading to breathing difficulties and lung infections.

In bulbar palsy, the throat muscles are principally involved, and difficulties in swallowing and speech may be the main features. Patients with speech difficulties have a typically "quacking" voice.

Sometimes, the muscles can be seen to twitch (fasciculation), and pain and stiffness can develop around any joint where the muscles are affected.

As the disease progresses, help with breathing is often needed. A tracheostomy is also sometimes carried out to make breathing easier. This involves putting a tube through a surgical opening into the trachea (windpipe) at the front of the neck. Despite these measures, chest infections and pneumonia often cause complications, and can be difficult to treat. Most people who have MND will die from it within five years

There are some notable exceptions,

however. Stephen Hawking, the physicist, noticed his first symptoms - clumsiness in his hands - over 40 years ago.

Diagnosis

There is no single test for MND. The diagnosis is usually made on the basis of symptoms and what a neurologist (a doctor specialising in conditions of the nervous system) finds when examining the nervous system. These will rule out other conditions, such as multiple sclerosis (MS).

Tests to help diagnose MND include:

- an electromyogram (EMG), where the muscles are electrically stimulated and their strength measured,
- a muscle biopsy, which involves removing a small piece of muscle under local anaesthetic, and examining it under a microscope.

Treatment

Riluzole (Rilutek) is the only drug currently available to treat ALS in the UK, and it only slows the progress of the disease by a couple of months. The National Institute for Clinical Excellence, which advises the NHS on new or expensive treatments, has recommended the drug for the ALS form of the disease.

New treatments under investigation include drugs based on antioxidants to "mop up" waste molecules before they damage nerve cells. Creatine, a

chemical involved in the distribution of energy within cells, and oxandrolone, an anabolic steroid that helps to maintain body weight and muscle mass, are also being explored.

Support

It's important that people with MND have enough information to help them understand their illness and plan for disability. Physiotherapists, occupational therapists and speech therapists can all offer advice and support. Relaxation and breathing exercises regularly will help to keep stress levels under control.

The support of family and friends is invaluable. Just being there and helping the person with their inevitable bouts of anxiety and stress may be just as important as any help that qualified professionals can give.

Voluntary organisations can be useful

sources of information about equipment to make the lives of people with MND easier and more comfortable. They can also provide practical advice about financial issues such as state benefits.

It often helps to talk to others in a similar situation, and voluntary organisations may also be able to put people in touch with other MND patients.

Further information

Motor Neurone Disease Association

☎ 08457 626 262

www.mndassociation.org

Scottish Motor Neurone Disease Association

☎ 0141 945 1077

www.scotmnd.org.uk

Contact a family

☎ 0808 808 3555

www.cafamily.org.uk