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Motor neurone disease (ALS)

Motor neurone disease (MND) is a neurological condition causes a progressive weakness and wasting of many of the muscles in the body. There are various types of motor neurone disease. This leaflet is mainly about amyotrophic lateral sclerosis (ALS), which is the most common type of motor neurone disease. Although there is no cure for motor neurone disease, treatments can help to ease symptoms and disability.

What is motor neurone disease?

In motor neurone disease (MND), motor nerves become damaged and eventually stop working. Therefore, the muscles that the damaged nerves supply gradually lose their strength.

There are various subtypes of MND. In each type, symptoms tend to start in different ways. However, as the disease progresses, the symptoms of each type of MND tend to overlap. This means that symptoms in the later stages of each type of MND become similar.

Types of motor neurone disease

The main types of MND are:

Amyotrophic lateral sclerosis (ALS)

Amyotrophic lateral sclerosis (ALS) is the most common form of MND. About 8 in 10 people with MND have this type. Symptoms tend to start in the hands and feet. The muscles tend to become stiff as well as weak at first. ALS is also sometimes known as Lou Gehrig's disease in the United States of America, after the baseball player who was diagnosed with it.

Progressive bulbar palsy (PBP)

About 2 in 10 people with MND have this type. The muscles first affected are those used for talking, chewing and swallowing (the bulbar muscles).

Progressive muscular atrophy (PMA)

This is an uncommon form of MND. The small muscles of the hands and feet are usually first affected but the muscles are not stiff.

Primary lateral sclerosis (PLS)

This is a rare type of MND. It mainly causes weakness in the leg muscles. Some people with this type may also develop clumsiness in the hands or develop speech problems.

The rest of this leaflet is just about the ALS type of MND (ALS-MND).

Motor neurone disease (ALS) symptoms

This disease affects the muscles in different ways.

The main feature of ALS-MND is muscle weakness which is mild at first, but gradually becomes worse. Usually, the first symptoms of ALS develop in the hands and arms or in the feet and legs. Less commonly, the first symptoms are in the muscles around the face and throat (the bulbar muscles):

Hand and arm symptoms

- Weakened grip.
- Dropping things.
- Difficulty opening bottles, turning keys, etc.
- Muscles in your hands (especially at the base of your thumbs) become flatter with time.

Feet and leg symptoms

- Dragging one leg.
- Tripping easily and more frequently.
- Difficulty climbing stairs.
- Difficulty getting up out of low chairs.
- Tiredness after walking.

Bulbar muscle symptoms

- Inability to shout or sing.
- Slurred speech.
- Changes in the quality of your voice.
- Difficulties with swallowing as the muscles which co-ordinate swallowing become affected.

Other symptoms
Other that may occur include:

- Muscle cramps.
- Tiredness.
- Twitching of weakened muscles (fasciculation).
- Jerking of an arm or leg whilst you rest.

How do ALS symptoms progress?

Symptoms may affect just one arm or leg at first. They then gradually become worse and spread to involve other limbs. ALS eventually causes many muscles in the body to become affected. Some muscle groups may be more badly affected than others. Muscles gradually become weaker and wasted. The problems that develop may eventually include the following:

- Walking (mobility) typically becomes worse.
- Doing tasks with your arms and hands becomes increasingly difficult.
- Eating, drinking and swallowing become difficult when the tongue and the muscles around the mouth and throat become weak.
- Sneezing and coughing become weak.
- You may become breathless on minimal exertion when the chest muscles become affected.

What causes motor neurone disease (ALS)?

The cause is not known. It is thought that certain chemicals or structures that only occur in motor nerves are damaged in some way. The reason why the nerves become damaged is not clear. (It is also not clear why sensory nerves, which have a similar structure, are not affected.)

There may be one or more environmental factors that may trigger the damage in someone who is prone to the disease. Research continues in order to find the underlying cause of the damage to the nerves.

How common is motor neurone disease (ALS)?

ALS motor neurone disease is uncommon in the UK. There are about 4,000 people in the UK with ALS-MND.

Who gets motor neurone disease (ALS)?

ALS-MND can affect anyone. ALS-MND is rare under the age of 40 years. It usually develops between the ages of 50 and 70 years. It is more common in men than in women.

For the majority of cases, it is not inherited and it does not run in families. In around 1 to 2 out of 20 cases of ALS, a family member has been affected, and there may be an inherited factor involved.

What is not affected in motor neurone disease (ALS)?

- **Intellect does not usually change**, as the thinking part or the brain is not usually affected. However, in about 10-15 out of 100 people with MND, a type of dementia develops. In up to 35 out of 100 others, there may be a mild effect on thinking ability.
- **Sensory parts of the nervous system are not affected**, so you can feel, see, smell, taste and hear as before.
- **Bladder and bowel functions usually remain normal**, so incontinence is not usually a feature. However, poor mobility may cause continence problems in the later stages of the illness.

- **Emotional feelings and sexual desire** are not directly changed by ALS-MND. However, some people with ALS-MND suddenly cry or laugh in inappropriate moments. (This is not a sign of mental illness but a feature of ALS-MND and cannot be helped, although may be embarrassing when it occurs.) Also, depression and anxiety are common.

How is motor neurone disease (ALS) diagnosed?

Unfortunately, there is no test that confirms ALS-MND. People with suspected ALS-MND are normally referred to a specialist (neurologist) for assessment. At first it may be difficult for a specialist to be sure that you have ALS-MND when early minor symptoms first develop.

However, the diagnosis usually becomes clear as the typical symptoms and signs of the disease gradually become worse. It can typically take a year from when the first symptoms start to the diagnosis being confirmed.

Tests are often done to exclude other causes of your symptoms. The following tests may be advised:

- **Electromyography (EMG)**. In this test, very small needles are used to record the nerve impulses occurring within your muscles (usually in your arms, legs and throat).
- **Nerve conduction studies**. An electrical impulse is applied through a small pad on the skin to measure the speed at which your nerves carry electrical signals.
- **Transcranial magnetic stimulation (TMS)**. This test measures the activity of the nerves which run from your brain to your spinal cord.
- Other tests, such as a magnetic resonance imaging (MRI) brain scan, may be done to rule out other causes of your symptoms if there is doubt about the diagnosis.

Motor neurone disease (ALS) treatment

Although there is no cure for ALS-MND, treatments can help both to slow the disease and also to improve any symptoms you may have.

Riluzole

Riluzole is a medicine which has been shown in clinical trials to have a beneficial effect on the survival of people with ALS-MND. It can slow down the disease progression by a few months. Riluzole works by its effect on glutamate - a chemical which relays signals between a nerve and another cell in the central nervous system.

Glutamate in excess has been shown to cause brain and spinal cord nerve damage. Riluzole helps to inhibit the amount of glutamate released in nerve impulse transmissions. This has some protective effect on the nerves.

Respiratory care

In this treatment, you are given a mask ventilator system to wear overnight while you are sleeping. The machines are small and portable and you can choose from a variety of different masks to suit you. People using this have been shown not only to have longer survival but also an improvement in the quality of their lives.

Treatments to help ease symptoms

Treatments are available for many of the symptoms that you may develop with ALS-MND. These may include cramps, difficulty swallowing saliva and food, urinary symptoms and depression.

It can be fairly common for people with ALS-MND to have feeding problems. These may either be due to difficulty swallowing or to difficulty in actually feeding yourself due to the weakness in your arms and hands. Some people find that having thickened fluid and sitting very upright when eating are beneficial.

If you are losing weight as a result of having feeding difficulties then you may benefit from having a gastrostomy inserted. This is a small feeding tube that is placed through the wall of the tummy (abdomen) directly into your stomach. Your doctor will discuss this in more detail with you if this might be appropriate for you.

Most people with ALS-MND are cared for by a professional team which includes neurologists, specialist nurses, physiotherapists, speech and language therapists, occupational therapists, dieticians and counsellors. Each problem that arises is assessed and dealt with as far as possible by the relevant members of the team.

What is the outlook for motor neurone disease?

MND does shorten life expectancy, and is a fatal illness. However, there is huge variation in how quickly ALS progresses, and it is different in each person who has it. Muscles weakened by ALS-MND do not recover. However, weeks or months may go by where the disease does not seem to progress. Eventually, severe disability develops. As the disease becomes severe, people with ALS-MND are unable to walk, talk or eat, and need a lot of care.

How quickly does motor neurone disease progress?

Your specialist may be able to give you an idea of how quickly MND is likely to progress in your particular case. However, the outlook (prognosis) for people with ALS-MND is extremely variable:

- About 7 in 10 people with ALS-MND die within three years of the onset of symptoms.
- About 25 in 100 survive five years.
- About 5-10 in 100 survive 10 years or more.

In the other, rarer types of MND, progress may be slower and the outlook better.

There are many research studies looking at different potential treatments for ALS-MND. It is hoped that new treatments will be introduced in the future.

Understanding nerves

Nerves (neurons) are like wires that carry tiny messages (electrical impulses) between the brain, the spinal cord and the rest of the body.

- **Motor nerves** carry messages from the brain and spinal cord to muscles, and make the muscles contract.
- **Sensory nerves** carry messages of touch, temperature, hearing, smell, taste and other sensations from various parts of the body to the brain.

Further reading

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