Medicine for Managers

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Motor Neurone Disease

Motor Neurone Disease (MND) is another of the unpleasant nervous diseases which are frightening and progressive. It causes nervous system damage which results in the muscles supplied by the nerves becoming weak and wasted.

In order to understand the disease it is necessary to understand how the nervous system works. Essentially there are two sorts of nerves; sensory nerves which carry electrical impulses (messages) from receptors for touch, pain, temperature, vibration and the special senses such as hearing, sight, smell, etc. **to** the brain, and motor nerves which carry electrical impulses (response messages) **from** the brain via the spinal cord to the muscles. For example, if you touch a hot plate, receptors in your hand will carry electrical impulses to the brain to register that your hand is touching something too hot. The brain interprets the messages and initiates a response which is carried by motor nerves in this example to the muscles of the arm, telling them to contract and pull the hand away from the hot plate. The motor nerves stimulate the muscles to work.

Loads of information is available from the Motor Neurone Disease Association
08456 626262 or
01604 250505
www.mndassociation.org
In Scotland
0141 945 1077
www.scotmnd.org.uk

In motor neurone disease the motor nerves degenerate and stop working. In consequence the muscles supplied by the affected nerves gradually become weaker and cease to work. Initially the disease may present with weakness in the hands resulting in loss of strength or dropping things, generalised feelings of tiredness and muscle cramps and sometimes pain. As the disease progresses the symptoms become more profound and debilitating until, ultimately, they cannot move the body.

Anyone can get MND but it is rare under 40 and most cases first develop between 50 and 70. It is nearly twice as common in men as in women. In about one in twenty people there is a family history of the disease. Overall about 2 people in 100,000 develop MND each year in England and there are about 5,000 people with the disease.

There are various classifications of MND. Broadly it is divided into three types.

- *Amyotrophic lateral sclerosis* (ALS) – affects about 8 in 10 sufferers and starts with weakness in the hands and feet.
- *Progressive Bulbar Palsy* – affects about 2 in 10 patients involving the mouth and throat causing chewing and swallowing difficulties and slurred speech.
- *Other atrophic disease* – may affect the lungs causing shortness of breath, or small muscles of the hands or weakness in the leg muscles. These forms are relatively rare.

The cause of MND is not clear. The reason why nerves are damaged largely remains a mystery. There is some evidence that a nervous system chemical messenger called glutamate may be involved in damaging the nerves. It could also be that there are environmental factors that could trigger the disease in someone who is susceptible.

The progress of the disease is insidious, particularly initially. Muscle weakness is mild and often starts with noticing that hand grip is less strong. Sufferers drop things and struggle to remove bottle tops. There may also be visible wasting in the muscles of the hands. One leg may start to be dragged or falls and trips may occur frequently. Climbing stairs may become more difficult and there is increased tiredness after walking. Sometimes difficulty shouting or singing may develop and speech becomes slurred. Voice quality may alter and difficulty swallowing may occur. Muscle cramps and twitching of muscles (fasciculation) may develop together with unexpected jerking of the limbs at rest. Over time, all the symptoms worsen and spread with some muscles more badly affected than others. Eating and swallowing become more difficult and coughing and sneezing become weaker. Tiredness may become more extreme and breathlessness increases, even on minimal exertion.

Usually intelligence and intellect is not affected by the disease (apart from very rarely when the disease is associated with dementia), sensations are normal, bladder and bowel activities are usually unaffected and emotion and sexual desire remains.

The diagnosis of MND is usually made from the history and by examination although various neurological tests may be undertaken to exclude other causes of the symptoms.

Sadly the outlook for MND is poor. Survival time is variable but is normally about 2-5 years, depending on the type of disease.

However, some people with the rarer types of disease can live for up to 10 years and some have lived for several decades, the most famous of whom is Professor Stephen Hawking, the British Physicist who was diagnosed with the disease over 40 years ago.

Management of patients with the disease is to provide supportive measures tailored to each individual's disabilities. This may involve adaptations to the house to aid mobility, speech therapy and feeding tubes and airway support to aid food intake and breathing. Oxygen may be required. The drug *Riluzole* is recommended by NICE for the most common form of MND (ALS) and should be initiated by a specialist although it can be managed by a GP. The drug, which is taken twice daily, is, however, very expensive at £320.00 for 56. It inhibits the quantity of glutamate released in nerve transmissions.

The end of life is usually precipitated by breathing difficulties. Most people pass away comfortably and painlessly in their sleep.