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MUSCULAR DYSTROPHY FOUNDATION • AUSTRALIA

JERRY LEWIS
International Patron

Facts About Motor Neurone Disease

What is Motor Neurone Disease?

Motor Neurone Disease (MND) is the name given to a group of closely related disorders affecting the motor neurones (nerve cells controlling muscles) in the brain and spinal cord. Degeneration of these motor neurones results in progressive muscle wasting and weakness because the muscles have lost their nerve supply. There are three main types of MND which are different forms of the same disease:

a) Amyotrophic Lateral Sclerosis (ALS)

b) Progressive Muscular Atrophy

c) Progressive Bulbar Palsy

There is considerable overlap between these clinical categories. ALS is the most common type and, indeed in the USA the name “ALS” is generally used to cover all types of MND. But in Australia the term MND is more commonly used.

If you have MND and are not sure which type you have, then your doctor will be able to help you.

What are the symptoms and effects of MND?

In the early stages of the disorders, the symptoms may be very slight. In addition, the disease may well have been present for some time prior to the onset of the symptoms. Early symptoms often consist of twitching (fasciculations) and cramps in the affected muscles and general fatigue, all of these symptoms sometimes being worse after exercise. Patients may also experience stiffness (spasticity) and jerking of the arms and legs.

The weakness, which occurs in MND usually, starts in the hands or feet and some muscles may be affected much more than others. Because MND is a progressive disease, the muscular weakness becomes worse with time. The patient may eventually require a wheelchair because of leg and trunk weakness, and become generally immobile.

At some stage in the disease the patient will suffer from “bulbar” symptoms. The bulbar area is located in the brainstem (lower part of the brain) and is important for production of speech and swallowing. Because there is loss of motor neurones in the bulbar region, there is difficulty with swallowing, chewing (due to tongue weakness), and speech. The MND patient may have “thick” speech, which can be slurred, nasal and monotonous.

Some patients with MND appear to have a loss of voluntary control over their emotions. He/she may suddenly laugh or cry inappropriately, and this may be distressing for both the patient and family. It is important for everyone to understand that these symptoms are part of the physical disease, and do not necessarily indicate a psychiatric problem. Although these symptoms (due to a “pseudobulbar palsy”) are well recognized, it is not fully understood why they occur. It should be emphasised that MND does not affect the senses (touch, taste, sight, smell and hearing) or the intellect, the patient will be able to think, reason and experience normal emotions at all stages of the disease. MND does not directly affect bowel, bladder or sexual function, although these may be indirectly affected later in the disease.

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In most cases, MND is steadily progressive and an average course is about four years. However, the disease may be very variable in length, and some patients have lived for 10-20 years. In a few cases progression of the disease has actually halted spontaneously, but this is rare. The reasons for the marked variability seen in MND are not yet fully understood. Many patients undoubtedly manage to live full, productive and satisfying lives during their illness, even when the disease is quite far advanced.

Who gets MND?

There is no evidence whatsoever that MND is “catching”, - it is not contagious. Most people who have MND are over the age of 40 years, but there have been cases as young as 20. Men are affected approximately twice as often as women. It should be appreciated that MND is not a rare disease, and approximately one person in 50,000 will develop MND in any one year.

In a very small proportion of cases (5-10%) there appears to be a type of familial pattern to the disease in which more than one member of a family is affected. However, in the great majority of MND cases, there is no hereditary pattern, and the children of affected individuals do not have an increased risk of developing the disease compared to the rest of the population.

MND occurs in countries all over the world, with about the same percentage of the population being affected. However, there are a few small areas of the world where MND occurs at an abnormally high rate. These include the Western Pacific island of Guam, Western New Guinea and the Japanese Kii Peninsula. The reasons for the unusually high occurrence of MND in these regions in not understood, and much research is currently being carried out to find the cause.

What causes MND?

No one yet knows the cause of MND. A number of possibilities have been explored in the past, and particular attention has been focused on a possible viral infection or environmental toxin as the cause. Although it is true that viruses do cause some neurological disorders, there is as yet no definite evidence that MND is caused by a virus.

How is MND diagnosed?

It is sometimes difficult to diagnose MND with certainty in the early stages of the disease. The diagnosis is usually made by a neurologist who will give the patient a thorough physical examination before arranging specialized tests. The most important investigation is the EMG (electromyogram) which involves measuring the electrical activity of muscles by means of a fine needle which is inserted into them. Muscles which have lost their nerve supply can be detected because their electrical activity is different from normal healthy muscles.

It is occasionally necessary to exclude pressure on the spinal cord as a cause of the symptoms, and for this reason the neurologist may order an MRI scan. Occasionally a sample of the spinal fluid is taken from the suspected MND patient.

It is not unusual for the patient with suspected MND to have various tests repeated at intervals of a few months, in order to permit a definite diagnosis. Sometimes these are done as an outpatient, but other patients are referred for hospital admission for these to be performed.

Is there a cure for MND?

We do not know what causes MND, and so we do not have any specific treatment which can reverse or slow the progress of the disease. No treatment that has been tried so far has had any effect on the course of the disease. However, many of the symptoms of MND can be treated, and this is of great importance to MND sufferers and their families.

The relief of symptoms can be discussed with staff of The Muscular Dystrophy Association.

Some notes on diet

Nutrition is an important consideration in the health care of any person and the person with MND is no exception. It is very important to continue to take a well-balanced diet - there are many unusual diets offered to patients, but as yet The Association has no evidence that they are of any benefit to patients at all.

Lack of knowledge about MND in the community may affect you too. Family and neighbours may not understand and may misinterpret the situation. Many families complain that in an effort to explain they describe the persons 'nerves' as being affected. This is then misconstrued by visitors as meaning that the patient is mentally ill, or, if speech is affected, brain damaged, and unable to understand. Both are hurtful to you and your family and may cause you to feel further isolated and alone. Professional workers with little knowledge of MND can unwittingly exacerbate this feeling by admitting their lack of knowledge - you may feel at best unique and at worst 'freakish'.

The Muscular Dystrophy Association's purpose and program

The Muscular Dystrophy Association (MDA) is committed to providing HOPE for people who suffer from the devastating nerve and muscle disorders. The only way this can be done is an all out offensive to find a control or cure for such disorders. MDA supports medical and scientific research to the extent that funds will allow, it runs a comprehensive public education program and provides physical aids, welfare, moral support and counselling to persons in need. The Muscular Dystrophy Association's programs are funded, almost entirely, by voluntary contributions from concerned individuals and community organisations.