

## ***Types of MND***

In Britain, amyotrophic lateral sclerosis (ALS) is the most frequent distinct type of MND. ALS can be further divided into a *familial* form (where the condition is inherited) and a *sporadic* form (where it is not inherited). Sporadic ALS is by far the most common form of MND, accounting for about 90% of all people with the condition. Progressive muscular atrophy, which affects only the lower motor neurones, is a less common form of MND, and causes deterioration more slowly.

Bulbar palsy is another form of MND, but this affects speech and swallowing. The nerve cells affected are located towards the back of the brain (the brain stem). These nerve cells are a specialized group that controls the cranial nerves to the muscles related to speech and swallowing.

### **What is the difference between ‘upper’ and ‘lower’ MND? My neurologist mentioned it as if it was very important.**

The important distinction is in the signs of the disease as revealed by the medical examination. This depends on whether the upper or lower motor neurones are mostly degenerated. Upper motor neurones are nerve cells that connect the brain to the spinal cord, extending downwards (in a collection of nerve fibres called the pyramidal tract); only a small number of upper motor neurones control many muscle fibres. Lower motor neurones connect the spinal cord to each individual muscle in the body so there are a much larger number of these. These two main types of nerve cells (neurones) are called ‘motor’ because they deal with movement of the muscles. The upper motor neurone meets the lower motor neurone in the front of the central part of the spinal cord (known as the anterior horn cell region).

Damage to a motor neurone will affect the muscles that it supplies with nerve impulses and possibly other nerves and muscles supplied further down the central nervous system (the brain and spinal cord). Therefore damage to a lumbar nerve might affect the lower limbs but could not affect muscles above this area. In

contrast, damage to an upper motor neurone (in the spinal cord or brain) may affect muscle groups in the head and arms as well as legs.

**My brother's son has had the diagnosis of MND, although he is only 6 years old. I thought it was found only in older people?**

There are two conditions that affect children and adolescents that are comparable but not the same as MND in adults.

The first is Werdnig–Hoffman disease, which is usually diagnosed at birth or within the first few months of life. Also called progressive spinal muscular atrophy, it usually results in death within the first few years of life. It affects about 1 in every 20,000 children born in the UK and accounts for 0.2% of all cases of motor neurone diseases.

The second is the very rare Kugelberg–Welander disease, also called chronic benign spinal muscular atrophy, which usually affects older children or adolescents. The outlook is one of slow (chronic) progression of muscle weakness with sometimes only limited disability. Kugelberg–Welander disease affects about half as many children as does Werdnig–Hoffman disease and accounts for about 0.1% of all cases of motor neurone diseases.

## ***Symptoms of MND***

**I have had a lot of weakness recently, and will be going soon to have tests to see if I have MND. Is this a significant symptom? Are there others?**

The first symptoms of MND indicating that something is wrong vary from the trivial to the severe. These symptoms can include foot-drop, where the lower leg muscles fail to lift the foot when walking (resulting in slapping the foot down with consequent trips and falls), difficulty in handling small objects and dropping things, slurred speech, extreme muscle tiredness, cramps and

muscle twitches (‘fasciculations’), and emotional instability (‘lability’), a symptom of uncontrollable laughter and crying without any obvious reason.

At the onset of MND, about 50% of sufferers will experience weakness of their arms, 25% weakness in legs or feet, while the rest have symptoms involving speech or swallowing.

Unlike many other neurological diagnoses, the mind, senses and bladder function are rarely affected in MND. This means that symptoms involving memory, thought processes (such as forgetfulness or confusion) or the senses (vision, smell, taste, hearing and touch) would be due to another disease.

Irrespective of the initial symptoms, MND will over time usually affect other limbs as well. The onset can be succeeded rapidly by further symptoms, so that many have quite significant impairments by the time they are diagnosed with MND; it is always important to have an assessment as early as possible after the condition is first considered. The box opposite indicates common and less common symptoms that can be expected – they are also common in lots of other conditions as well!

### **You talked about upper and lower MND earlier. What are the differences in symptoms?**

The symptoms of upper motor neurone degeneration include stiffness (spasticity); this is assessed by resistance to passive movement (increased ‘tone’). The tendon jerks assessed with a reflex hammer are exaggerated because the muscle contracts too strongly on testing. There is also widely spread, rather than localized, weakness. Because of the small number of upper motor neurones and because each upper motor neurone supplies signals to so many lower motor neurones, the symptoms are more noticeable because they will be more wide ranging.

The symptoms of lower motor neurone loss include marked weakness in individual muscles, causing partial or total paralysis with thinning of the muscles (‘wasting’). In these cases there is a decrease in, or absence of, muscle reflexes as the muscles fail to contract on testing with a reflex hammer. There are also irregular and involuntary contractions (‘fasciculations’). Lower motor

**Common symptoms**

- weakness
- tiredness
- depression, anxiety and insomnia
- constipation

**Less common**

- hunger pangs
- urinary symptoms
- pressure sores
- diarrhoea

neurone signs are generally more localized, affecting single muscles, although groups of muscles can also be affected.

Because lower motor neurones supply nervous impulses to a single muscle, or even a part of a muscle (in combination with other nerves signalling the rest of the same muscle), damage to a single lower motor neurone may be very limited or even imperceptible. In contrast, the effect of damage to upper motor neurones is more significant and widespread. Distinguishing between upper and lower signs can be difficult, as the initial symptoms such as weakness can be identical and both types of signs can be present; it is only by repeated assessment of symptoms that the degree of upper and lower motor neurone degeneration can be estimated.

**When I look closely I can see little ripples in my muscles and I get this terrible ‘creepy’ sensation as if something is walking up and down my leg. If this is caused by my MND, is it going to get worse? Can I do anything to stop it?**

The sensations you are experiencing are called ‘fasciculations’, irregular and involuntary contractions of your muscles. Because they are not voluntary but spontaneous and happening all by

themselves, they can be a disturbing experience, in the same way as finding that your arm or foot in bed has ‘gone to sleep’ and doesn’t feel as if it’s part of you. Some degree of this (usually occasional and slight) is fairly common in absolutely normal people and they do not indicate anything untoward. These contractions are harmless and painless but they can be extremely disturbing. When severe or continuous, they can be a sign of damage to the lower motor neurones.

In MND, fasciculations can become more frequent and noticeable but they do not cause pain or interfere with activity but may lessen when muscle wasting occurs.

If there is pain and difficulty in mobility, they can be helped by treatment. Stiffness and fasciculations can be helped to a greater or lesser degree with the muscle relaxants baclofen (Lioresal) or dantrolene (Dantrium); cramps can be lessened using the anti-malarial treatment quinine (the chemical that gives tonic water its distinct bitter flavour), the anti-anxiety drug diazepam (Valium), or the anti-epileptic drug phenytoin (Epanutin).

### **As MND is a neurological disease, is it going to affect my mental capacity eventually? How soon will it start to affect my mind?**

MND directly affects only the motor neurones (the nerve cells concerned with movement) – so it is unlikely that intellectual function will be noticeably affected. Many neurological conditions do have side effects of impaired thinking skills (‘cognition’), confusion or forgetfulness (impairment of short- or long-term memory); research has shown that those people with MND rarely have cognitive problems that are directly related.

Those with MND have psychiatric conditions such as depression, panic attacks and other problems in common with the population at large; as the disease progresses, these may become more frequent as a response to this deterioration.

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**My neurologist has said that my ‘mind may be spared’ in MND, but my occupational therapist has warned me about emotional problems – who is right?**

Strangely enough, both are correct. It is commonly said of MND that the senses (sight, touch, hearing, smell and taste) and brain function are spared – which is what your neurologist has told you. There are no psychiatric disturbances, mental illness or other mental problems associated with MND, so the severe forgetfulness or confusion that might be associated with other brain diseases are unlikely to affect people with MND.

Some people with MND find that they occasionally have emotional instability (‘lability’) when they laugh or cry for no apparent reason, or there is inappropriate or exaggerated emotional responses to situations, when they are unable to control these extreme feelings; this may be what the occupational therapist has warned against. Not all those with MND will experience emotional instability, since it occurs only in those who have upper motor neurone degeneration on both sides of the brain (see the next question on ‘pseudo-bulbar palsy’).

Depression and emotional lability must be clearly distinguished. Depression affects 30% of all adults at some time and should be treated in MND just as it should be in those without MND, with appropriate counselling and drug treatment. Emotional lability, in contrast, is a symptom associated with upper motor neurone degeneration but may be helped by medication.

**My husband has been diagnosed with bulbar palsy. What sort of symptoms would he expect to have with this?**

In about a quarter of all people with MND, the first symptoms at onset and diagnosis are ‘bulbar’ symptoms (the bulb being the hind brain) – these symptoms include slurred speech (‘dysarthria’) and difficulty in controlling swallowing (‘dysphagia’). Bulbar symptoms are not specific to MND and are more common in those who have had strokes on both sides of the brain.

There are in fact two types of bulbar palsy depending on whether the upper or lower motor neurones are affected on both

sides of the brain. Both types give difficulty in speech and swallowing but when the lower motor neurones are affected, the tongue will show thinning (‘wasting’) and fasciculation (called ‘fibrillation’) in the tongue; this type is called ‘chronic bulbar palsy’. Only in the upper neurone type is there emotional lability and this type is called ‘pseudo-bulbar palsy’.

**The overwhelming sensation I have is lack of strength; everything is such an effort. Why are my muscles so weak?**

Even in healthy people, when muscles are not used to their full capacity, the body stops using nutrients to maintain their size and volume. After some time, the muscles begin to lose mass (‘atrophy’) and become smaller and thinner. This wasting is much worse in MND.

Strength and muscle mass can be helped by correct diet and physiotherapy, but a full physical assessment is needed for a detailed plan, depending on the cause.