MOTOR NEURONE DISEASE

# When cells malfunction

*By Xena Fox*

**The term Motor neurone disease (MDN) is used to describe a group of neurological disorders that affect the way muscles work, such as muscular atrophy, primary lateral sclerosis, and progressive bulbar palsy.**

The most common is Amyotrophic lateral sclerosis (ALS), the condition often being synonymous with MDN. All are neurodegenerative and cause increasing disability which leads to eventual death. ALS became well known worldwide when physicist Stephen Hawking was diagnosed with it in 1963.

## What is ALS?

ALS may be the most common motor neurone condition, but it is still classified as a rare disease. In Europe and the United States, it affects around 2.2 people out of each 100,000.

Motor neurones are specialist nerve cells that pass messages to voluntary muscles telling them what to do. When they can no longer send instructions properly, the muscles cannot function correctly, leading to weakness, wasting and loss of control. Walking, sitting, eating, speaking and breathing can all be affected. It can be classified in several different ways: by where it starts, how symptoms progress, and whether it is inherited or not.

In approximately 70 per cent of cases, the limbs are affected first. In roughly a quarter, muscles in the face, mouth, and throat. Five per cent of sufferers find that muscles in the trunk of the body are first to be affected. In a few cases, symptoms can be limited to one region of the spine.

The earliest mention of the disease dates back to at least 1824. Fifty years later, the neurological connection was first described by French Neurologist Jean-Martin Charcot in an 1874 paper, from which the term amyotrophic lateral sclerosis originates as well. Sadly, though knowledge of the condition has advanced, there is still no known cure and understanding of its causes is limited.

***↑ Most people are diagnosed with ALS in their 50s, 60s or 70s***

***→ Sometimes symptoms are too vague to be worrying at first***

## Onset and progression

Symptoms generally appear when people are in their 50s or 60s. Nonetheless, ALS can affect adults of all ages, including teenagers (though rarely), but the majority of those diagnosed are over the age of 40. It affects slightly more men than women, though comparisons vary depending on the type. Percentages also even out after the age of about 70.

The initial symptoms may be so subtle that they are overlooked. They include slight clumsiness, mild weakness or difficulty speaking. It's easy

to blame them on tiredness or other reasons, so it's often some time before someone feels they should visit their doctor.

Symptoms build up gradually over weeks and months, usually on one side of the body. They may include a weakened grip, which can make picking up or holding objects difficult; awkwardness when walking or running; difficulty with tasks such as buttoning a shirt or writing; weakness at the shoulder that makes lifting the arm difficult; cramping; slurred, very quiet, or nasal speech; loss of tongue mobility; and difficulty swallowing.

Symptoms depend on the type of ALS. They also vary enormously from person to person, as does the pattern of their progression. Nevertheless, over time, people will have increasing mobility difficulties, and will experience other problems such as stiffness, exaggerated reflexes and muscle wastage. As damage