Australian Brain Alliance

CRACK THE BRAIN'S CODE

MOTOR NEURONE DISEASE

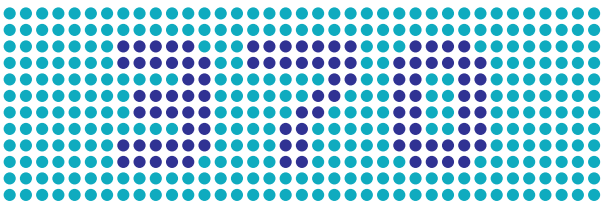
What is motor neurone disease?

Motor neurone disease occurs when neurons within the motor cortex region of the brain (upper motor neurons) and spinal cord (lower motor neurons) degenerate and die.

These upper and lower motor neurons are responsible for motor function—a person's ability to move, speak, breathe, walk or run. Without stimulation from the neurons (nerves) they are connected to, muscles around the body become weaker and waste away.

People with the condition have trouble walking, breathing, swallowing, difficulty with speech, or holding objects. They can also suffer from cramps or muscle twitching. Malnutrition and respiratory failure are major problems that result from motor neurone disease.

Around 370 people are diagnosed with motor neurone disease each year in Australia.



Motor neurone disease can affect the upper motor neurons only, the lower motor neurons only, or both. It does not affect the sensory neurons—a person's ability to see, touch, smell, hear and taste is not affected. However, the condition can cause changes in the frontal or temporal lobes of the brain, which affect a person's language, behaviour, personality and cognitive ability.

Although some symptoms of motor neurone disease can be treated, it is still incurable. It is a progressive degenerative condition. Most people with the condition live for 1–5 years after being diagnosed.

Around 2000 people in Australia have motor neurone disease.

In a disease as relentlessly progressive and degenerative as motor neurone disease, early diagnosis is crucial.

While the exact causes of motor neurone disease are unknown, it appears to be a combination of genetic and molecular factors. Around 5–10 % of cases are attributed to genetic causes.

High levels of particular proteins are found associated with the neurons that suffer damage and die in motor neurone disease. This has led to a theory that the disease could in part be caused by abnormalities in the production and function of various proteins and their role in binding and transcribing certain genes. Cell damage and death can also occur as a result excitotoxicity, caused by high levels of glutamate, a common chemical in the brain used to send signals between neurons.

In a disease as relentlessly progressive and degenerative as motor neurone disease, early diagnosis is crucial. The presentation of symptoms can vary widely from person to person, and many of the symptoms are common to other conditions. Misdiagnosis, or delayed diagnosis can be significant issues in the identification of motor neurone disease. Advances in neuroimaging, including improved diffusion tensor MRI, functional imaging and network analysis have led to more effective diagnosis of motor neurone disease, primarily in enabling the exclusion of other possibilities.

As well as improved neuroimaging applications, transcranial magnetic stimulation (TMS) applications have also helped improve diagnostic capabilities. This technique involves non-invasive application of a magnetic pulse that induces an electrical current within the brain. This current can be targeted to stimulate particular regions or brain circuits. TMS can be used to pinpoint areas of upper motor neuron dysfunction in patients, and has been shown to speed up diagnosis by up to 8 months.

The variable nature of motor neurone disease also makes drug development difficult, as large-scale clinical trials are needed to properly test new drugs. The only therapeutic treatment available in Australia is a drug called riluzole. If prescribed early, riluzole can be effective in slowing the disease progression. Overall, it has been found to prolong median survival by two to three months.

Researchers at the University of Melbourne have recently embarked on a clinical trial to test a drug that delivers a drug containing a compound with neuro-protective properties, which was shown to delay the onset of symptoms in mice .

Other work being conducted at the University of Sydney is attempting to identify the underlying causes and mechanisms of motor neurone disease. Better understanding of these processes will help inform more effective treatments.

The Australian Brain Initiative will nurture the basic brain research required to better understand the complexities of conditions like motor neurone disease. The Initiative will also progress collaboration between research and industry to advance neurotechnology devices that have the potential to transform the capabilities of neuroscience research and improve diagnostic capabilities that will potentially help buy precious time for those living with motor neurone disease

Around 400 000 people worldwide have motor neurone disease.

Motor neurone disease is responsible for nearly 400 deaths per year in Australia.