New stem cell technology reveals fresh insights into motor neuron disease

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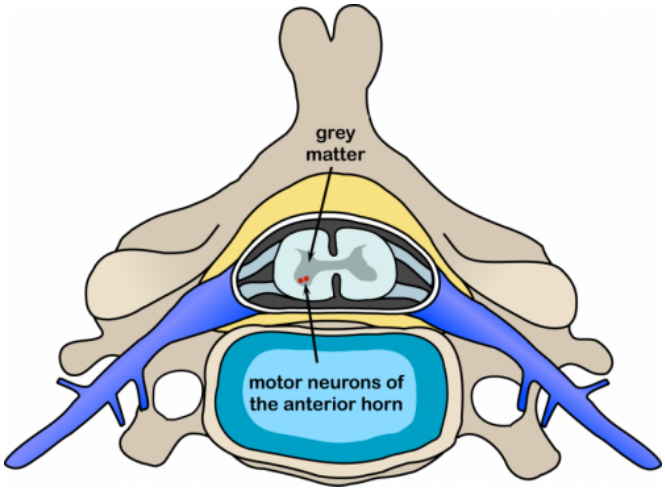

Polio spinal diagram. The poliovirus affects the motor neurons of the anterior horn cells, or the ventral (front) grey matter section in the spinal column, which control movement of the trunk and limb muscles including the intercostal muscles. Credit: Wikipedia/CC BY-SA 3.0

Scientists at the University of St Andrews have discovered new ways of studying what happens to motor neurons affected by motor neuron disease (MND) by using stem cells derived from patient skin samples, according to research published in *Nature Communications* today.

MND is a progressive neurodegenerative condition that attacks motor neurons, specialised nerve cells in the brain and spinal cord, causing the loss of signals from the brain to muscles and eventually leading to paralysis.

Approximately 5000 people in the UK live with MND at any one time, with approximately 5 people dying of the disease every day. There is currently no cure for MND and treatment options are limited.

The joint research project led by scientists from the University of St Andrews and the University of Edinburgh has shown that even before they show any signs of damage, motor neurons affected by MND lose the ability to generate the electrical signals required to make muscles contract due to changes in specialised proteins called ion channels.

Dr Gareth Miles from the University of St Andrews and lead researcher on the project, said:

"Learning more about how and why motor neurons are lost in MND plays a crucial role in developing new treatments and ultimately finding a cure for this devastating disease. Using new developments in stem cell technology has enabled us to compare the function of motor neurons from healthy individuals with those from patients suffering from different forms of MND."

Dr Miles was supported in his research by University of St Andrews' PhD student Anna-Claire Devlin and Professor Siddharthan Chandran from the University of Edinburgh.

Dr Miles continued:

"Our findings suggest this may be an early step in understanding and ultimately treating the disease process of MND and highlights ion channels as potential targets for future therapies. Our work also demonstrates that studying the function of stem cell-derived motor neurons could be important for the development and testing of new drugs to treat and eventually cure the disease."

About motor neurone disease (MND):

- MND is a fatal, rapidly progressive disease that affects the brain and spinal cord.
- It attacks the nerves that control movement; people can still think and feel, but their muscles refuse to work.
- It can leave people locked in a failing body,

unable to move, talk, and eventually, breathe.
- It affects people from all communities.
- It kills five people every day in the UK - 30% within a year and more than 50% within 2 years of diagnosis
- It has no cure.

More information: Anna-Claire Devlin, Karen Burr, Shyamanga Borooah, Joshua D. Foster, Elaine M. Cleary, Imbisaat Geti, Ludovic Vallier, Christopher E. Shaw, Siddharthan Chandran and Gareth B. Miles. "Human iPSC-derived motoneurons harbouring TARDBP or C9ORF72 ALS mutations are dysfunctional despite maintaining viability." (in press)

Provided by University of St Andrews