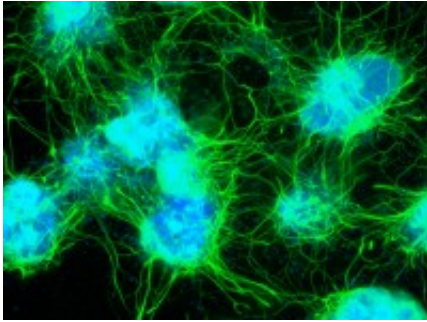


Study gives clues to causes of motor neurone disease

10 October 2012



The team used neurones derived from embryonic stem cells for the study.

(Medical Xpress)—Scientists at the University of Bath are one step further to understanding the role of one of the proteins that causes the neurodegenerative disorder, Amyotrophic Lateral Sclerosis (ALS), also known as Motor Neurone Disease (MND).

The scientists studied a protein called angiogenin, which is present in the spinal cord and brain that protects neurones from cell death. Mutations in this protein have been found in sufferers of MND and are thought to play a key role in the progression of the condition.

MND triggers [progressive weakness](#), [muscle atrophy](#) and muscle twitches and spasms. The disease affects around 5000 people in the UK.

The team of [cell biologists](#) and structural biologists have, for the first time, produced images of the 3D structures of 11 mutant versions of angiogenin to see how the mutations changed the structure of the active part of the molecule, damaging its function.

The study, published in the prestigious journal *Nature Communications*, provides insights into the causes of this disease and related conditions such

as Parkinson's Disease.

The team also looked at the effects of the malfunctioning proteins on neurones grown from [embryonic stem cells](#) in the laboratory.

They found that some of the mutations stopped the protein being transported to the [cell nucleus](#), a process that is critical for the protein to function correctly.

The mutations also prevented the cells from producing stress granules, the neurone's natural defence from stress caused by low [oxygen levels](#).

Dr Vasanta Subramanian, Reader in Biology & Biochemistry at the University, said:

"This study is exciting because it's the first time we've directly linked the structure of these faulty proteins with their effects in the cell.

"We've worked alongside Professor Ravi Acharya's group to combine structural knowledge with cell biology to gain new insights into the causes of this devastating disease.

"We hope that the scientific community can use this new knowledge to help design new drugs that will bind selectively to the defective protein to protect the body from its damaging effects."

The findings were welcomed by medical research charity, the [Motor Neurone Disease](#) (MND) Association, the only national charity in England, Wales and Northern Ireland dedicated to supporting people living with MND while funding and promoting cutting-edge global research to bring about a world free of the disease.

Dr Brian Dickie, Director of Research Development at the charity, said: "The researchers at the University of Bath have skilfully combined aspects of biology, chemistry and physics to answer some

fundamental questions on how angiogenin can damage motor neurones. It not only advances our understanding of the disease, but may also give rise to new ideas on treatment development."

More information:

[www.nature.com/ncomms/journal/ ...
full/ncomms2126.html](http://www.nature.com/ncomms/journal/full/ncomms2126.html)

Provided by University of Bath

APA citation: Study gives clues to causes of motor neurone disease (2012, October 10) retrieved 8 October 2022 from <https://medicalxpress.com/news/2012-10-clues-motor-neurone-disease.html>

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