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Stem cell study homes in on ALS cause

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By Maggie Fox, Health and Science Editor

WASHINGTON (Reuters) - Mutated nerve cells called glial cells may secrete the poisons that cause amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, researchers reported on Sunday.

Two reports published in the journal Nature Neuroscience may show new ways to treat the degenerative nerve disease, which slowly paralyzes its victims until they die.

Both studies used embryonic stem cells from mice to generate batches of cells that mimicked the disease. The researchers said the studies demonstrate that embryonic stem cells can be vital for basic medical research.

Such batches of cells could also be used to test new drugs to treat the incurable and almost always ALS, also known as motor neuron disease.

"These findings are particularly significant for two reasons," said Dr. Kevin Eggan, a stem cell expert at Harvard University who led one of the studies.

"They provide a proof of concept -- if you have embryonic stem cells that carry the genes for a disease, in this case ALS, you can make limitless quantities of the cells effected by the disease (and) study the disease process," he said in a statement.

"Additionally, both we and our colleagues at Columbia have demonstrated that in patients with this particular genetic form of ALS there is a toxic factor causing the cells to die."

Both teams showed that the nerve cells called astrocytes, which are supposed to support and feed neurons, turn toxic when they carry a mutated gene called SOD1, which has been linked with ALS in the past.

Dr. Serge Przedborski of Columbia University in New York and colleagues created mouse motor neuron cells that carried mutated versions of the human superoxide dismutase-1 or SOD1 gene. But these mutated cells did not cause the damage typically seen in ALS when grown in lab dishes of cells.

Then they created astrocytes carrying the mutated human SOD1 gene. Astrocytes are one of the types of glial cells -- support cells in the brain and nervous system that secrete various compounds that nourish neurons.

When SOD1 is mutated in these glial cells, Przedborski and colleagues found, one of the nourishing proteins apparently turns toxic. When they grew astrocytes with mutated SOD1, they killed the neighboring mouse motor neuron cells.

"These findings indicate that astrocytes may play a role in the specific degeneration of spinal motor neurons in ALS," Przedborski's team wrote.

If that particular bad protein can be identified, it might lead to a drug that could treat ALS.

The use of embryonic stem cells can provide a good way to test potential treatments, both teams of researchers said.

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