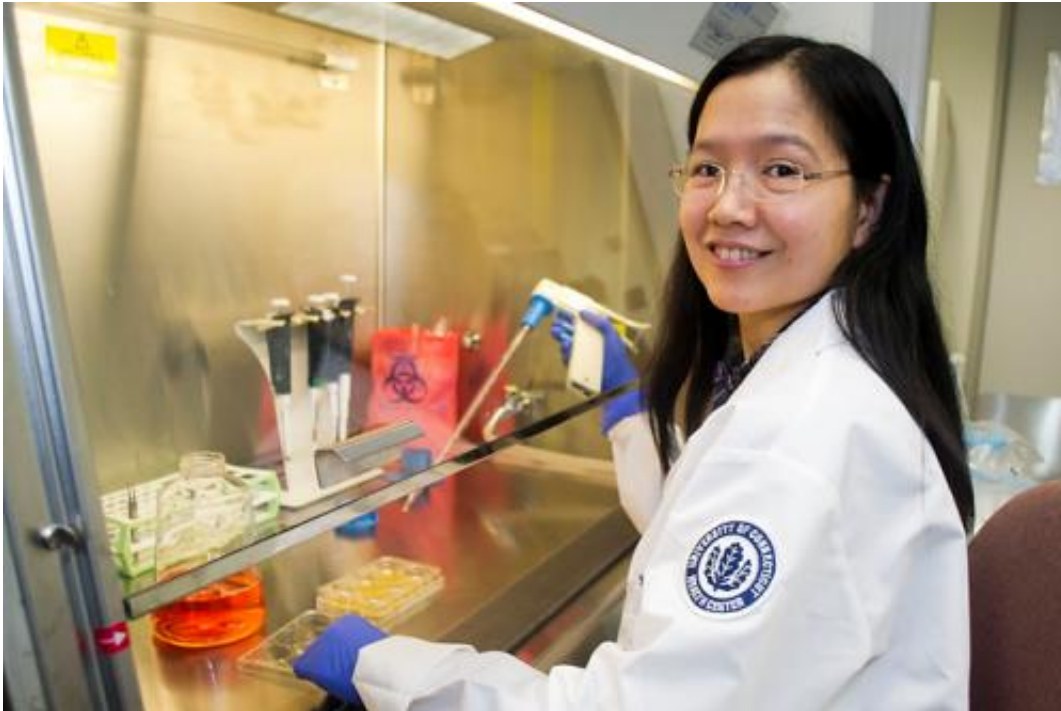


# Researcher advancing motor neuron studies

January 29 2013, by Carolyn Pennington

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Xue-Jun Li, Department of Neuroscience, is using stem cells to study the devastating condition known as spinal muscular atrophy. Credit: Tina Encarnacion/UConn Health Center Photo

A University of Connecticut researcher is advancing the understanding of the devastating inherited condition known as spinal muscular atrophy.

Xue-Jun Li, assistant professor in the Department of Neuroscience, is corresponding author of a paper published in the prestigious journal *Cell Research* in December 2012 entitled "Recapitulation of spinal motor

neuron-specific [disease phenotypes](#) in a human cell model of spinal muscular atrophy." The paper's other authors are UConn Health Center researcher Zhi-Bo Wang and Xiaoqing Zhang of the Tongji University School of Medicine in Shanghai.

Spinal [muscular atrophy](#) (SMA) is a group of inherited diseases that cause [muscle damage](#) and debilitation, which progress over time and eventually lead to death. To be affected, a person must inherit the [defective gene](#) from both parents. About 1 in 10,000 people have SMA, and most do not survive childhood due to [respiratory problems](#), [heart failure](#) and infections.

"There is no effective treatment for [spinal muscular atrophy](#), and one of the roadblocks is not knowing why the spinal motor neuron degenerates," Li explains. "One of the aspects of our research is to understand how specific types of neurons are specified and degenerated. We are trying to model neurological disorders by using human motor neurons derived from [stem cells](#)."

Establishing human cell models of SMA to mimic motor neuron-specific phenotypes holds the key to understanding this destructive disease, she says. The model described in the journal article provides a unique paradigm for studying how motor neurons degenerate. It also highlights the potential importance of antioxidants for the treatment of SMA.

Understanding how [motor neurons](#) are specifically degenerated can lead to effective interventions in the future. "It can help us find some way to rescue the motor neuron degeneration in this disease," Li points out. "Understanding the role of antioxidants can provide potential clues to finding a treatment."

Li's interest in medicine began in high school when she considered a career as a physician, but she refocused on medical research as a college

undergraduate in Shanghai. She describes her cousin's battle with a neurological disease as the trigger for her concentration in that field. "It made me really want to find some cures and understand the disease," she says. "I wanted to do something that improves general health."

Li moved to the U.S. in 2002 and worked for five years as a researcher at the University of Wisconsin in Madison. She was drawn to the University of Connecticut because of the state's stem cell research grant program and the university's creation of a Stem Cell Institute. Now there are even more opportunities with the commitment of state leaders to support genomics and personalized medicine, and The Jackson Laboratory construction of a major research facility on the Health Center's campus.

"That's why UConn was so attractive to me," she says. "There was strong support for my research. I applied for a grant from the state and got it right away. I feel very lucky to be here."

Provided by University of Connecticut

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