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THEME: 3 B Nervous system Série S

Potential New Approach for Improving Quality of Life for Amyotrophic Lateral Sclerosis Patients

The disease starts slow. It begins with minor missteps, small errors typing. But within a matter of only a few years, those small mistakes, falls and fumbles¹ progress to complete paralysis. The muscles that keep you breathing stop doing their job, leading to lethal respiratory failure. And the whole time, cognition remains intact, rendering you keenly aware of what is happening, helpless to improve the condition.

Lead investigator Steven Burden, and colleagues show that, by increasing the signaling activity of a protein called MuSK, they were able to keep nerve cells attached to muscle longer into the progression of the disease in a mouse model of ALS. The withdrawal, or detachment, of motor nerve terminals from muscle is the first sign of disease in all forms of ALS, followed by the death of nerve cells, or neurons. When the terminals detach, the muscles are denervated, or disconnected from the nerve, leaving the neurons no longer able to send messages to muscles, and the ability of the brain to initiate and control muscle movement is lost. Vast research efforts have focused on ways to keep the neurons from dying...

The neuromuscular synapse is required not only to move, but also to breathe and live." "It is always exciting and gratifying when findings from fundamental, basic science can be applied to treat disease," Dr. Burden said. "When we started (in 1993), we discovered MuSK in fish and could only speculate about its function. Then MuSK was found in other vertebrates, including mice and humans, and we learned that it is essential to form and maintain neuromuscular. It's very satisfying to study a molecule and a process in detail, to understand how a molecule works, how a synapse forms and then to apply this information in a disease setting. We don't have all the answers and we don't know with absolute certainty that our approach will work in humans, but we're confident that we're headed in the right direction. Fortunately, we have a mouse model of ALS that replicates the human disease, so we are hopeful that increasing MuSK activity in ALS patients will likewise preserve attachment of nerve terminals and improve motor function."

From ScienceDaily (Aug. 30, 2012)

Sum up this article and explain the main ideas using your scientific knowledge

¹ To fumble : mishandle, mismanage