Preserving nerve cells in motor neuron disease

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A team of researchers, led by Scott Oakes, at the University of California, San Francisco, has identified a way to prevent symptom onset, weight loss, and paralysis and extend survival in a mouse model of amyotrophic lateral sclerosis (ALS; also known as Lou Gehrig's disease), providing a new avenue of research for the development of therapeutics for ALS and other motor neuron diseases.

ALS and other motor neuron diseases are neurological disorders that selectively affect nerve cells that control voluntary muscle activities such as speaking, walking, breathing, swallowing, and general movement of the body. A key feature of these diseases is that the affected nerve cells (which are known as motor neurons) die by a process known as apoptosis.

Determining whether this death contributes to disease or occurs after the nerves have stopped functioning is important to establishing whether blocking apoptosis would have therapeutic benefit. In the study, genetically eliminating activation of the mitochondrial apoptotic pathway in a mouse model of ALS was shown to preserve motor neuron viability and function, thereby preventing symptom onset, weight loss, and paralysis and extending survival.

The authors therefore suggest that inhibiting activation of the mitochondrial apoptotic pathway might provide a way to preserve motor neurons in individuals with ALS and other motor neuron diseases.