

Seminar in Neuroscience



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**8th Floor
Neuroscience Center
of Excellence
Conference Room**
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Why Corticospinal Motor Neurons Are Important For ALS and Other Motor Neuron Diseases

Corticospinal motor neurons (CSMN) are some of the most important cortical components of motor neuron circuitry. Their unique ability to collect, integrate, translate and transmit the brain's input to the spinal cord targets allow them to function as the spokesperson for the cerebral cortex for the initiation and modulation of voluntary movement. CSMN vulnerability and progressive degeneration is key in numerous motor neuron diseases, such as primary lateral sclerosis, hereditary spastic paraplegia, and amyotrophic lateral sclerosis. CSMN death also leads to long-term paralysis in spinal cord injury patients. Therefore, it is important to understand the cellular and molecular mechanisms that are responsible for the vulnerability and degeneration of this clinically-relevant neuron population. Since their numbers are very limited and there are no cellular markers to label them, investigation of CSMN biology has been challenging. We developed novel tools and approaches to make these neurons visible *in vivo*, and characterized a model system in which CSMN display progressive degeneration. Recently we identified ER-stress to be one of the underlying cellular mechanisms that leads to their degeneration, which becomes evident by apical dendrite degeneration and spine loss. Understanding the biology behind CSMN degeneration will help develop effective treatment strategies for many motor neuron diseases in which voluntary movement is impaired.