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Amyotrophic Lateral Sclerosis (ALS)

Amyotrophic lateral sclerosis (ALS) is a mixed lower motor neuron and upper motor neuron disorder. Another name for ALS is Lou Gehrig's disease. ALS affects only motor neurons. However, some of these motor pathways can be in the autonomic nervous system. Amyotrophic in the name of this disease comes from the word roots "a" (absent), "myo" (muscle) and "trophy" (nourish). Together, amyotrophic refers to the muscle weakness that develops in ALS.

ALS affects LMNs located in the anterior horn of the spinal cord, motor nuclei of the brainstem, and UMNs with cell bodies in the precentral gyrus of the frontal lobe. Atrophy and shrinkage of skeletal muscles is common in ALS due to the loss of UMNs that descend the spinal cord and the resulting inability to stimulate and move muscles. Ocular motility and the parasympathetic neurons in the sacral spinal cord are spared. Specimens of the spinal cord from individuals who had ALS show degeneration of lateral columns of white matter in the corticospinal tracts. Hence the term "lateral sclerosis" found in the name.

ALS affects men more often with the typical age of onset being late adulthood (60 or older). Typically, patients only survive 2-5 years after the onset of symptoms. Early symptoms of ALS include asymmetric spasticity, cramping, and weakness of the arms and legs. Patients may initially drop objects or have difficulty with fine motor skills like typing, writing, or using eating utensils. As the disease progresses, more muscles are affected and all limbs become involved. Eventually, muscles of the head become affected and patients experience mastication impairment, dysphagia (which increases the risk for aspiration), and dysarthria (slurred speech) due to lost control of muscles. Respiratory musculature is affected later in the course of the disease. Respiratory failure is often the cause of death for ALS patients.

The cause of ALS is still largely unknown. A mutation in a gene that codes for superoxide dismutase (SOD1) was originally thought to impair the ability to rid the body of free radicals. Now scientists think ALS may be caused by mutated SOD1 proteins that fold incorrectly and form aggregates that are toxic to neurons or trigger apoptosis. Treatments are being considered that may help clear these misfolded aggregates.

There is no cure for ALS, so treatment involves managing symptoms and efforts to slow the progression of the disease. The only approved drug for this disease is riluzole (Rilutek). It acts to reduce levels of glutamate in order to slow neuronal destruction.