

Public Abstract

First Name:Brandie

Middle Name:Renee

Last Name:Morgan

Adviser's First Name:Martin

Adviser's Last Name:Katz

Co-Adviser's First Name:Joan

Co-Adviser's Last Name:Coates

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Title:Motor Unit Characterization in Canine Degenerative Myelopathy: A Disease Model for Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a fatal neurodegenerative disease. Clinical onset occurs in individuals ~40 years and older, and patients experience progressive muscle paralysis. Death typically occurs 2-5 years after clinical onset due to paralysis of muscles involved in breathing. There is no effective treatment for ALS, and there is a critical need for the identification of disease models to assist in treatment development. In this study, we worked to characterize a potential dog model of ALS known as canine degenerative myelopathy (DM). DM is a fatal naturally occurring neurodegenerative disease found in many breeds, has a disease progression very similar to ALS, and is associated with mutations in a gene also linked to human ALS. Muscle and spinal cord tissue from dogs affected with DM were assessed for motor unit (MU) (motor neuron, motor axon, and muscle fiber) death, from a region of the spinal cord involved in facilitating breathing. Our results indicate that functional impairment occurs before physical degeneration of MU structures. Our findings of degeneration in sensory neurons and axons suggest that DM is a disease affecting motor and sensory systems. Sensory abnormalities have been reported in some forms of ALS. The work conducted in this study has contributed to our overall understanding of DM, has further highlighted similarities between ALS and DM, and has strengthened the notion that DM is a good disease model for some forms of ALS. Upon further characterization of DM, future studies will utilize this model to develop effective treatments for ALS patients, and DM affected dogs.