

# MOTOR UNIT CHARACTERIZATION IN CANINE DEGENERATIVE MYELOPATHY: A DISEASE MODEL FOR AMYOTROPHIC LATERAL SCLEROSIS

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## ABSTRACT

Canine Degenerative Myelopathy (DM) is a late onset, heritable neurodegenerative disease that occurs in many breeds. DM clinical signs and disease progression are comparable to some forms of amyotrophic lateral sclerosis (ALS). As with some forms of ALS, DM is associated with mutations in superoxide dismutase 1 (*SOD1*), indicating that DM may be a potential ALS disease model. DM has only recently been identified to affect motor units (MUs) (motor neuron, axon, and myofibers), which are structures commonly affected in ALS. Thus, in an effort to further characterize DM as an ALS disease model, the focus of this study was to further explore MU involvement in the DM disease progression. Results from this investigation indicate that thoracic intercostal muscle atrophy in DM is not preceded by physical loss of the motor neurons innervating these muscles, or of their axons. Axonal loss in thoracic sensory roots and sensory nerve death suggest that sensory involvement may play an important role in DM disease progression. Results from this study support previous findings that DM is a multisystem disease with many similarities to human familial *SOD1*-D90A ALS, indicating that DM is a suitable disease model for this form of ALS. Further analysis of the mechanisms responsible for these morphological findings would aid in the development of therapeutic intervention for both diseases.