

VETERINARY TECHNICAL DATASHEET

Acral Mutilation Syndrome (AMS)



Mutation Found In :French Spaniel, German Shorthaired Pointer, Pointer, English Springer Spaniel

Disorder Type

- Nervous system

Disease Severity

- Moderate/severe

Background

Acral Mutilation Syndrome is a severe hereditary sensory neuropathy. The disorder is characterized by loss of pain sensation in the paws that causes paw ulceration and self-mutilation. This disorder is caused by the abnormal development and progressive degeneration of sensory neurons. Motor neurons are not affected by this disorder.

Key Signs

- Excessive licking or biting of paws

Clinical Description

Clinical signs of the disease typically emerge at around four months of age. One or more paws of affected dogs become unable to feel pain. Affected dogs tend to lick or bite their paws excessively. This self-mutilation is severe and often results in toenail loss, fractures, and digit amputation. The motor abilities and proprioception of affected dogs remain normal. It has been suspected that the trigger for paw biting and licking in these dogs are minor paw injuries caused by insensitivity to pain. The disorder can be suspected based on breed and typical clinical signs. The neurological status of affected dogs is normal except for their paws lacking sensory capacity.

Mode of Inheritance

- autosomal recessive

Gene Name

- GDNF

Next Steps

There is no curative treatment for the disorder. Affected dogs are treated according to their condition and the severity of clinical signs. Affected dogs must be closely monitored so that infection and paw trauma can be spotted as early as possible. Self-mutilation must be avoided. The disorder is severe and usually leads to euthanasia on welfare grounds.

References

Plassais J, Lagoutte L, Correard S, Paradis M, Guagure E, He ´dan B, Pommier A, Botharel N, Cadiergues MC, Pilorge P, Silversides D, Bizot M, Samuels M, Arnan C, Johnson R, Hitte C, Salbert G, Me ´reau A, Quignon P, Derrien T, Andre C. A Point Mutation in a lincRNA Upstream of GDNF Is Associated to a Canine Insensitivity to Pain: A Spontaneous Model for Human Sensory Neuropathies. PLOS Genetics | DOI:10.1371/journal.pgen.1006482 December 29, 2016.