VETERINARY TECHNICAL DATASHEET

Globoid Cell Leukodystrophy or Krabbe Disease, (GLD); mutation originally found in Terriers

Mutation Found In :Cairn Terrier, West Highland White Terrier

Disorder Type

- Neuromuscular

Disease Severity

- Severe

Background

Globoid cell leukodystrophy is a neurologic lysosomal storage disease that causes severe neuronal degeneration. The underlying cause of the disorder is a deficiency of a lysosomal galactocerebrosidase enzyme. This enzyme deficiency causes defects in the production of myelin that protects and insulates neurons. This particular genetic variant has been found to cause GLD in West Highland White Terriers and Cairn Terriers.

Key Signs

- Muscle weakness
- Ataxia
- Tremor
- Paralysis
- Behavioral changes

Clinical Description

GLD is characterized by muscle weakness, tremors, and ataxia (uncoordinated movement). Clinical signs of the disease also include behavioral changes, incoherence, blindness, and deficits in normal reflexes. GLD is a progressive condition in which the characteristic signs of the disease are first observed in the hind legs in dogs 1 to 5 months of age.

Mode of Inheritance

- autosomal recessive

Gene Name

- GALC

Next Steps

The wellbeing of affected dogs should be monitored carefully. Treatment is supportive care and symptomatic depending on the severity of the dog's clinical signs. Affected dogs do not typically survive to adulthood and are usually euthanized on welfare grounds before 9 months of age.

References

Victoria T, Rafi MA, Wenger DA. Cloning of the canine GALC cDNA and identification of the mutation causing globoid cell leukodystrophy in West Highland White and Cairn Terriers. Genomics 33:457-462, 1996.