VETERINARY TECHNICAL DATASHEET

Spinocerebellar Ataxia with Myokymia and/or Seizures, (SAMS)

Mutation Found In :Russell Terrier, Parson Russell Terrier, Fox Terrier (Smooth), Fox Terrier (Toy)

Disorder Type

- Nervous system

Disease Severity

- Moderate/severe

Background

Early onset spinocerebellar ataxia is a neurological disorder characterized by uncoordinated movements and impaired balance. The mutation has been found in various terrier breeds and is inherited in an autosomal recessive fashion.

Key Signs

- Ataxia
- Hypermetria
- Impaired balance

Clinical Description

Clinical signs of this condition are usually first detected when the puppy is between 2 to 6 months of age. The first observable sign of spinocerebellar ataxia is lack of muscle coordination, particularly evident in the pelvic limbs. They may also exhibit hypermetria, myokymia (muscle fasciculations), neuromyotonia (muscle twitching at rest), excessive facial rubbing, and seizures. Affected dogs are often euthanized due to difficulties walking.

Mode of Inheritance

- autosomal recessive

Gene Name

- KCNJ10

Next Steps

Depending on the severity of the clinical signs, a dog can be supported to reduce the likelihood of injury when moving around. Stairs may pose a particular risk and should be avoided. Medical control of seizures can be attempted. However, humane euthanasia may be elected.

References

Gilliam D, O'Brien DP, Coates JR, Johnson GS, Johnson GC, Mhlanga-Mutangadura T, Hansen L, Taylor JF, Schnabel RD. A homozygous KCNJ10 mutation in Jack Russell Terriers and related breeds with spinocerebellar ataxia with myokymia, seizures, or both. J Vet Intern Med. 2014 May-Jun;28(3):871-7.

Rohdin C, Gilliam D, O'Leary CA, O'Brien DP, Coates JR, Johnson GS, Jäderlund KH. A KCNJ10 mutation previously identified in the Russell group of terriers also occurs in Smooth-Haired Fox Terriers with hereditary ataxia and in related breeds. Acta Vet Scand. 2015 May 23;57:26.