

Spinocerebellar Ataxia (SCA)

Affected breeds: Parson Russell Terrier, Jack Russell Terrier

Spinocerebellar Ataxia is a progressive condition in which the affected dog loses coordination and balance when moving. The onset of the condition is typically at 2 – 10 months of age. In many cases affected dogs also suffer from myokymia (involuntary muscle twitching) and/or epileptic seizures. SCA affected dogs are usually euthanised.



Another test for ataxia in the PRT and JRT, known as LOA (Late Onset Ataxia), is also available. There are also as yet unidentified mutations which cause other forms of ataxia in these breeds.

Spinocerebellar Ataxia is caused by a recessive genetic mutation. This means that dogs which carry the mutation ("CARRIERS") are normal but will pass the mutation on to an average of 50% of their offspring. Puppies which inherit two copies of the mutation are at very high risk of Spinocerebellar Ataxia ("AFFECTED").

This test is particularly useful for breeders:

- To identify carriers among their breeding stock so that they can avoid CARRIER X CARRIER mating combinations which would risk AFFECTED puppies.
- To conclusively confirm Spinocerebellar Ataxia in an affected dog

This test will be reported as:

CLEAR : no evidence of the Spinocerebellar Ataxia mutation

CARRIER : carries one copy of the defect, which will be passed to 50% of offspring

AFFECTED : carries two copies of the defect, and are at very high risk of SCA

The genetic status of dogs can be used to predict breeding outcomes when different combinations are mated:

CLEAR X CLEAR = 100% CLEAR

CARRIER X CLEAR = 50% CARRIER, 50% CLEAR

AFFECTED X CLEAR = 100% CARRIER

CARRIER X CARRIER = 25% AFFECTED, 50% CARRIER, 25% CLEAR

AFFECTED X CARRIER = 50% AFFECTED, 50% CARRIER

AFFECTED X AFFECTED = 100% AFFECTED

References

D Gilliam, DP O'Brien, JR Coates, GS Johnson, GC Johnson, T Mhlanga-Mutangadura, L Hansen, JF Taylor, and RD Schnabel (2014) A Homozygous *KCNJ10* Mutation in Jack Russell Terriers and Related Breeds with Spinocerebellar Ataxia with Myokymia, Seizures, or Both. J Vet Intern Med 28:871-877