Juvenile-onset Laryngeal Paralysis and Polyneuropathy (JLPP*)

*Recently re-named Polyneuropathy with Ocular Abnormalities and Neuronal Vacuolation (POANV) in the RBT and referred to as Neuronal Vacuolation and Spinocerebellar Degeneration (NVSD) in the Rottweiler

Affected breeds: Russian Black Terrier, Rottweiler

Juvenile-onset Laryngeal Paralysis and Polyneuropathy (JLPP) is an inherited condition in which the nervous system deteriorates. Initially the nerve controlling the larynx (the voice box) is affected, resulting in noisy breathing and problems swallowing. Later the nerves controlling the hind-legs are affected, making it progressively more difficult for the dog to get up and stand without wobbling. Symptoms typically start at a few weeks of age; euthanasia is indicated due to a poor quality of life.

JLPP 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 will develop JLPP ("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 JLPP in an affected dog

This test will be reported as:

CLEAR : no evidence of the JLPP mutation
CARRIER : carries one copy of the defect, which will be passed to 50% of offspring
AFFECTED : carries two copies of the defect, causing JLPP

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

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