

CEREBELLAR ATAXIA 5

A BRIEF SUMMARY OF WHAT WE KNOW SO FAR

CEREBELLAR ATAXIA IN THE ITALIAN SPINONE

(Pending the issue of an information pamphlet to be written by Dr. Bruce Cattanach)

1. Although this disease has not yet been fully identified, or named, it has been made quite clear, by Dr. Simon Wheeler, Senior Lecturer in Neurology at The Royal Veterinary College, that this is a new neurological disease affecting the cerebellum. This part of the brain provides the fine tuning to movement and control of gait. The affected dogs show exaggerated movements such as a high stepping gait of goose stepping and also an unsteady or wobbly gait, generally referred to as ataxia. There is no treatment for this disease and the puppies are put to sleep when they are no longer able to cope with the symptoms, usually at around 11 - 12 months of age.
2. CA only affects young dogs, all the cases reported so far (13 puppies out of 7 litters in the UK and 3 puppies out of 2 litters in Denmark, born between November 1990 and March 1997) have been under one year old. Therefore, even though an adult dog may be a descendant of one or more of the proven carriers it is highly unlikely that it will develop CA, although it could be a carrier.
3. The clinical signs of CA are as described in a separate report.
4. CA appears to be restricted to one family line only and all cases reported so far have one or more of the proven carriers on both sides of the pedigree.
5. From the above information, and by studying the pedigrees, together with the number of affected puppies in the litters, it has been shown that the mode of inheritance of CA is that of a simple recessive gene i.e.
 - a) for affected puppies to be produced both the sire and the dam must be carriers.
 - b) it is the unidentifiable carriers which spread the disease throughout the breed (the puppies which are affected never reach the age when they could be bred from and therefore pose no threat).

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c) a brief example of the possible mating's are shown below:

i) a carrier mated to a carrier will produce puppies of the following status:

25% clear ----- 25% affected ----- 50% carriers
Therefore, 75% will appear normal.

ii) a carrier mated to a clear will produce puppies of the following status:

50% clear ----- 0% affected ----- 50% carriers
Therefore, 100% will appear normal.

iii) a clear mated to a clear will produce puppies of the following status
100% clear

Therefore, all will appear normal.

6. a) With a policy of out crossing it is possible to prevent the production of affected puppies and, if this were continued over a number of generations, the frequency of carriers being produced would decrease. The probably of descendants of proven carriers being carriers themselves would, on average, be reduced by a half in each subsequent generation e.g.

1st generation ----- 50% ----- or 1 in 2 risk of being a carrier
2nd generation ----- 25% ----- or 1 in 4 risk of being a carrier
3rd generation ----- 12.5% ----- or 1 in 8 risk of being a carrier
4th generation ----- 6.25% ----- or 1 in 16 risk of being a carrier
5th generation ----- 3.13% ----- or 1 in 32 risk of being a carrier

b) Under the guidance of geneticist, Dr. Bruce Cattanach, the Committee of the ISCSGB have formulated a control scheme to help prevent the production of affected puppies and also to reduce the frequency of production of carriers within the breed.

Please remember that if you have any questions, no matter how trivial they may seem, do not hesitate to contact me. You can ring me evenings or weekends on 01522 792743 (INT 44 1522 792743) or you can write to me at the following address:

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UPDATE: Another case of CA has been reported. This was a puppy bitch from an accidental grandmother to grandson mating. The bitch was actually 14 months old when she was put to sleep but I understand that the symptoms were very advanced and Dr. Wheeler had said it was the worst case he has seen so far, so one can only assume that the bitch had been affected for some time. This brings the total so far to 14 puppies from 8 litters in the UK and 3 puppies from 2 litters in Denmark.

Issued by Pat Wilkinson (Health Sub Committee - Ataxia) ISCGB 6/18/98