

# CEREBELLAR ATAXIA 3

## THE STORY OF CHIANTI AND TEABAG

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There have recently been two more cases of cerebellar ataxia reported, the two affected puppies being litter sisters. There is no treatment for this, as yet, unidentified disease, (the cerebellum is one of the structures of the brain and ataxia is usually defined as an unsteady or irregular gait resulting from lack of coordination of the muscles, which can be caused by a variety of lesions throughout the nervous system. In short, brain damage.) and the puppies are put to sleep, usually at around 10 - 11 months old.

First indications of the disease are clumsiness, the puppy does not pick its feet up properly and sways, sometimes mistaken for hip dysplasia. This is very noticeable when the puppy rises from a prone position and it may lean against furniture or walls in order to steady itself. At this stage the puppies always seem very much improved outdoors, being able to walk and trot much more freely and being able to keep their balance.

As the disease progresses the puppy loses coordination of the hind limbs, lifting the legs in a highly exaggerated manner and this progresses into the front limbs. The puppy finds it very difficult to move in a straight line and will sometimes lose its balance and fall over. However, the puppies always seem to find the strength to pick themselves up and will play normally with their companions. They also eat normally and retain their happy personalities to the end. It is most distressing to watch the progression of this disease, as I have recently with a puppy bred and owned by Christine Bennett, and sired by my own dog. The only saving grace is that there appears to be no pain associated with the disease.

Now follows a brief history of these two puppy bitches, from the day they were diagnosed as suffering from this disease, and also the steps being taken by the ISCGB to control the spread of the disease and ultimately to eradicate it from the breed.

12/29/97 Both bitches, Chianti and Teabag, having been referred by their own vet were diagnosed at a veterinary hospital as suffering from a form of Cerebellar Ataxia. Both had already undergone various tests to rule out distemper, toxacara and neospora. Both puppies had spinal fluid drawn from the base of the skull; this showed abnormalities.

(In late 1995, early 1996, a puppy had been referred to Dr. Simon Wheeler, a lecturer in neurology at the University of London, Royal Veterinary College. The puppy was owned by Peter and Linda Morris and bred by Chris Jackson. Chris and Linda took the puppy, together with an unaffected littermate and their dam, to Dr. Wheeler for further investigation. C.A. had already been diagnosed but Dr. Wheeler had offered to do some research to try to identify the disease. A video was made of the puppy and various tests carried out. When he was put to sleep tissue samples were taken from the brain and spinal cord but with very limited results. No further progress was made at the time despite the fact that this puppy's litter brother also had tissue samples removed and these were also sent to the RVC.)

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Dr. Simon Wheeler was contacted about Chianti and Teabag and an appointment was arranged for him to see Teabag.

1/16/98 Christine Bennett and myself took Teabag to the RVC where Dr. Wheeler took a note of her history, made a physical examination and video and also carried out electrophysiological tests as he believed that the nerves themselves were affected, not just the brain. He thought it may be a type of motor neurone disease and also said that it was more than likely hereditary. Christine had already agreed that when the time came she would donate Teabag's body for further research in order to help identify the disease.

2/4/98 Chianti died suddenly and, although her owners were devastated, they agreed to a post mortem and that tissue samples should be sent to the RVC to help with the research.

2/16/98 Teabag was put to sleep as it was becoming an effort for her to move around and she could only manage very short walks. Tissue samples were sent to Dr. Wheeler at the RVC.

2/20/98 I spoke to Dr. Bruce Cattanach, a well known geneticist, who agreed to help us.  
2/23/98 A copy of Teabag's pedigree, together with six others in which affected puppies had been confirmed as suffering from C.A. (dating from 1990-1995), were sent to Dr. Cattanach for his opinion on the possibility of the disease being hereditary.

Dr. Cattanach has since stated that we are clearly dealing with an hereditary disease, the mode of inheritance being that of a simple recessive gene. This is backed up by the number of puppies in each litter, the number of puppies showing the clinical symptoms and the relationship of the dogs in the pedigrees.

Dr. Wheeler is continuing his research and we are hopeful of some early news.

I must point out that both Dr. Wheeler and Dr. Cattanach are dealing directly with the ISCGB, with myself as the contact, and with no other persons, other than Teabag's owner, Christine Bennett. My aim in writing this report is to raise awareness of the disease with Spinone owners in the USA and to ask for their help in reporting any cases of ataxia, even if only suspected, to myself as committee member of the ISCGB so that we can work together in identifying the disease and eradicating it.

The frightening thing is that carriers of the disease will appear perfectly normal, showing no clinical signs whatsoever but, if they are mated to another carrier, again showing no signs of the disease, 25% of the puppies will be affected by the disease, 25% will be clear but the remaining 50% will be "silent" carriers. Hence, the disease spreads through the breed and people may be unwittingly mating carriers to clear dogs, quite unaware that they are producing more carriers.

The ISCGB will shortly be publishing a list of the known carriers, any dogs proven to be clear and also probable carriers, the last category being the most worrying. Our aim is obviously to assist Dr. Wheeler in his research and to work toward identifying the disease, the gene which carries it and to find the marker in the dog's DNA which will

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help to develop a blood test, but this is not going to happen overnight and we are busy raising funds to finance the research, etc.

We are relying heavily on people's honesty in reporting all cases of CA in puppies they have bred and/or owned. If they do not, then any scheme we introduce for the control of the disease will be flawed. The more information we can collect, the sooner we will be rid of this dreadful disease.

If anyone would like to speak to me directly they are quite welcome to ring me on INT 44 1522 792743.

Christine Bennett and myself are quite happy for anyone to see the pedigree of this latest litter, she being the breeder and me being the owner of the sire.

Pat Wilkinson

ISCGB Committee Member March 10, 1998