

## Neuropathy Update!

As you know, we are gathering information on the progressive neurological movement problem that is occurring in older solid colored English Cocker. Onset is typically seen between 8-10 years of age. It begins with a lack of coordination (ataxia) in the hind limbs. Affected dogs will wobble, knuckle over, or drag the rear feet. As the disease progresses the dog will buckle over and have difficulty standing and getting up. Eventually the dog is a paraplegic. This progressive weakness also affects the front limbs, facial muscles, and the ability to swallow. There is no pain. At this point we know there is a loss of nerve fibers in the spinal column that interferes with communication between the brain and limbs. Our disease process and clinical symptoms appeared to be similar to **D**egenerative **M**yelopathy. The human equivalent is Lou Gehrig's disease (**A**myotrophic **L**ateral **S**clerosis). Research has recently shown affected English Cocker are "unlikely to have a mutation in SOD1 or other genes that cause human ALS."

Currently, research on this devastating disease is being conducted at the University of Pennsylvania under the direction of Charles H. Vite, DVM, PhD. His team consists of Dr. Natalia Kunze, Pierce Nathanson, and Dr. Alice ChenPlotkin.

Sincere thanks are extended to those that have submitted blood samples and tissue for this research. We urge those that have an affected dog, or a dog that fits the above description to contact a Health Committee member for further information, or Dr. Vite at the University of Pennsylvania. His contact information is below.

This disease is relatively uncommon, but it has been diagnosed not only in North America, but other parts of the world. It is very important that those with affected dogs participate in this research. Dr. Vite has full support of the ECSCA. Help us to help our beloved breed.

Addi Pittman, Chairman  
Liz Neff  
Genelle Joseph  
Joyce Winkels  
Donna O'Connell  
Bruce Barrett, DVM

My name is Pierce Nathanson and I am a veterinary and Ph.D. student at the University of Pennsylvania. I have always had an interest in neurodegeneration and when I joined the lab of Dr. Alice Chen-Plotkin, a neurologist studying the genetics of neurodegenerative diseases in humans, I felt it was important to use my veterinary training to investigate not only neurodegenerative diseases that affect humans but those that affect animals as well. Indeed, in speaking with Dr. Charles Vite, a veterinary neurologist and researcher, I learned of a group of English Cocker Spaniels (ECS) with a previously unrecognized spinal cord disorder affecting their ability to walk and ultimately leading to hindlimb paralysis. It is my hope that with the participation of English Cocker Spaniel owners and breeders that I will be able to uncover the genetic cause of this disease.

To date we have generously received blood samples from English Cocker Spaniel owners with dogs that are both healthy and unaffected. Given the clinical similarity to dogs with degenerative myelopathy, our first goal was to ascertain if these dogs possessed mutations in SOD1—the gene that is the cause of degenerative myelopathy in several canine breeds as well as a subset of humans with Amyotrophic Lateral Sclerosis (ALS). Our preliminary results indicate that these dogs are unlikely to have a mutation in SOD1 or several other genes that can cause human ALS. Thus, the search continues for the cause of this devastating disease.

The more samples we are able to collect, the more likely we will be able to find the genetic mutation that causes this disease. We would appreciate any blood samples, pedigree information, videos, or post mortem samples of affected English Cocker Spaniels or unaffected English Cocker Spaniels over the age of 10 years. With the help of the ECS community, we hope to identify the genetic cause of the disease. This will allow us to design a genetic test so that owners and breeders will have a better understanding of the health of their beloved companions. Questions on sample submission may be directed to Dr. Charles Vite at the University of Pennsylvania School of Veterinary Medicine:

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