

Ataxia in Scotties: Wobbly Dogs  
by Carole Fry Owen  
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Murphy, a beautiful, healthy 12-week-old Scottie pup, came to me in 1994. He was typically full of energy and lightheartedness. He played hard and acted as any other puppy would. Nothing unusual was noticed concerning his movements, but in time I would learn that Murphy's future would be anything but normal. The burden he now carries is continual and without end. I need to tell his story because other Scotties are at risk, and even though nothing can help Murphy, maybe bringing his problem in the open will one day help others avoid the disease he is afflicted with. Also, finding a diagnosis for Murphy was a long and difficult process; others shouldn't have to wait the six years I did to finally have an answer."

Scottie Scamper, September 2000, author Debbie Smith

Murphy has cerebellar abiotrophy, and Debbie Smith, Harborcreek, PA, opened his story in the Canadian Scottish Terrier Club quarterly with the above paragraph. Debbie had tried to find out what was wrong with Murphy since he was seven months old. "Luxating patellas," the first veterinarian told her. Patellar (knee) surgery at two and a half years didn't make Murphy normal. In fact, his movement became increasingly uncoordinated. Later, veterinarians couldn't agree whether Murphy had a neurological problem or a neuromuscular disorder. When he was six, veterinarians finally pronounced Murphy to have some sort of progressive neurologic disease, but visits to assorted veterinarians had not narrowed the diagnostic possibilities.

All these years Murphy's condition had continued to worsen. Debbie's family carried Murphy up and down steps. Outside they supported him by a harness because he was very unsteady on his feet. Murphy would fall often and bang his chin. Eating and drinking were difficult because of his head bobbing and swaying. Today Murphy requires constant help, even to eat and drink. Otherwise, he topples over.

Debbie and husband Bill loved Murphy and tended to his needs, but they did not learn what caused his problem until recently. She had suspected for quite some time that Murphy had cerebellar abiotrophy (CA) as described in Gordon Setters, but couldn't get her veterinarian to agree because she could find nothing published on CA in Scotties. Debbie read an article in CSTC's The Scamper that put her on the track to answering her suspicions. Editor Leona Carter had reprinted "Health Search, Health Share" from The Bagpiper (V99:#2). The section about cerebellar abiotrophy caught Debbie's eye. It told of geneticist Dr. Jerold Bell's work on CA in the Old English Sheepdog. The article also mentioned statistics from the 1995 STCA Health Survey that show ataxia (a physical finding of CA) has a "guesstimated" carrier incidence of 13% in our breed.

What exactly is ataxia? Webster's Dictionary defines "ataxia" as "loss of coordination of the muscles, especially of the extremities." Very simply, an ataxic dog is wobbly. Murphy was and is ataxic.

Once Debbie knew of Dr. Bell's work with CA, she contacted him and then Dr. Alexander deLahunta, a renowned authority on cerebellar disorders at Cornell University in New York. Dr. deLahunta and Dr. Bell confirm that Murphy has cerebellar abiotrophy. Their diagnosis, based on physical findings, will be checked by autopsy when Murphy dies. Dogs with CA have cerebellums smaller than normal, at least in progressed disease, and certain abnormalities in brain cells. An MRI has already established that Murphy's cerebellum is small. CA-affected dogs aren't born with a smaller cerebellum though. It becomes smaller upon death of certain brain cells. Therefore, dogs diagnosed with CA early in life may not show a smaller cerebellum

While a trustee of STCA's Health Trust Fund, I learned through Debbie of Dr. Bell's interest in ataxia and cerebellar abiotrophy in Scotties. Dr. Jerold Bell is director of the clinical genetics course at Tufts University School of Veterinary Medicine and advises many national breed clubs on genetic issues. I wrote Dr. Bell for information.

Dr. Bell replied: "Cerebellar abiotrophy (ataxia)...is a slowly progressive degenerative condition that affects muscular coordination and movement. It does not affect the senses. Ataxia refers to the clinical symptom of incoordination, while abiotrophy refers to the premature cell death in the cerebellar region of the brain that causes the incoordination. This disorder is hereditary, and I have studied it in many breeds, including Gordon Setters, Old English Sheepdogs, Spinoni, and a related disorder in Kerry Blue Terriers. In these breeds, the disorder is inherited as a simple autosomal recessive trait...Some owners or even veterinarians may confuse the disorder with Scottie Cramp on first presentation. However, CA is a slowly progressive condition that does not go away, and Scottie Cramp is an episodic condition..."

"If the Scottish Terrier Club of America or the STCA Health Trust is interested in determining the mode of inheritance, objective frequency estimates, genetic spread of the defective gene(s) causing CA, and recommendations for genetic control, I am available to do so...if the STCA is interested in a study to determine the genetic parameters of cerebellar ataxia in the breed, it will require interviews of owners/breeders, veterinary confirmation of diagnoses, pedigree analysis, and informational releases to the club. Depending on the number of affected dogs, the cost for such a study would be up to \$1,000."

For Health Trust Fund to consider Dr. Bell's offer, it will be helpful to know more about incidence of cerebellar ataxia, and more specifically cerebellar abiotrophy, in Scotties. Although owners may have been told their Scotties have "progressive neurologic disease," it is impossible to know from the STCA's 1995 Survey if the ataxia reported is caused by cerebellar abiotrophy or other neurologic conditions.

To that end, I welcome calls from Scottish Terrier owners who have dogs with ataxic symptoms, i.e., dogs that are uncoordinated and "wobble." Any wishes for confidentiality will be honored. Owners or breeders may contact Carole Owen, 1203 N. 9th St., Temple, TX 76501; ph. 254/791-1365; e-mail to [ritescot@aol.com](mailto:ritescot@aol.com).

Owners who are uncertain what ataxia (or cerebellar abiotrophy) looks like may borrow a video Debbie Smith has compiled of her Murphy from 2 1/2 to 6 years of age. It shows the progression of Murphy's disease. I've seen the video, and it is inspiring to see the happy Murphy that resides in a dog body that doesn't work like other Scotties'. Murphy's video and a copy of Debbie's article "Cerebellar Abiotrophy (Ataxia) in the Scottie" from CSTC's The Scamper are available from Debbie Smith, 6556 Pinar Road, Harborcreek, PA 16421; ph. 814/899-2856; e-mail to Litlblkdog@aol.com.

"The disorder can be prevented in other Scotties when we understand the nature of the genetic process that resulted in this degeneration," Dr. deLahunta told Debbie Smith. "This is where Dr. Bell can be so helpful for you and the breed." (Author's note: When I contacted Dr. deLahunta, he mentioned being willing to do brain necropsy studies at no cost upon death of ataxic Scotties suspected to have cerebellar abiotrophy, as long as specimens are sent to him. For information, contact Dr. Alexander deLahunta, Department of Biomedical Sciences, College of Veterinary Medicine, Cornell University, Ithaca, NY 14853; ph. 607/253-3547; e-mail to ad43@cornell.edu.