



Chihuahua

Update

Vol. 10, No. 1 ■ May 2011

Chihuahuas Are Among Toy Breeds Affected by Syringomyelia

When a 4-year-old gold and white Long-Coat Chihuahua, GB/LUX CH Deeruss Flashmoon at Ballybroke, collapsed from muscle weakness and an inability to balance on his legs in December 2006, his co-owners and longtime Chihuahua enthusiasts were baffled. "Flash" represented excellent breed type and was a prominent stud dog.

"I bred Flash earlier that day, and he was fine," says Darwin Delaney of Dartan Chihuahuas in Essexville, Mich. "The next thing I knew, he was down."

Delaney took Flash to the veterinary teaching hospital at Michigan State University, where magnetic resonance imaging (MRI) indicated Flash suffered from syringomyelia (SM), a neurological condition in which fluid accumulates in the spinal cord, forming cavities called syrinxes.

Flash was treated with medications, including prednisone, an antibiotic and pain relievers. During the next three weeks, Flash recovered with rest. A couple of months later, co-owner Graham Foote of Ballybroke Chihuahuas in West Sussex, England, arrived to take Flash home with him. "The only evidence of his condition was a slight weakness in his right front leg," Foote recalls. "I continued giving him prednisone for about two weeks, but since then he has not needed medications."

Though Flash remains asymptomatic, a second MRI in 2008 confirmed the initial diagnosis. Now, nearly five years

later, Flash lives a normal life. Earlier this year, Flash won his class at the Crufts Dog Show in Birmingham, England. "Occasionally, Flash shows signs of stiffness in his right shoulder, but otherwise he remains in good health," Foote says.

Meanwhile, Delaney, who was concerned about potentially spreading the disease, neutered and spayed all of Flash's offspring.

Syringomyelia is a disorder of the central nervous system that commonly occurs in Cavalier King Charles Spaniels, with a 46 percent prevalence rate that is even higher in older dogs. Most Cavaliers are believed to have SM secondary to Chiari-like malformation (CM), a condition in which the brain is too big for the skull, causing it to be crushed and pushed out of the foramen magnum, the funnel-like opening to the vertebral canal. This obstructs the flow of cerebrospinal fluid in and out of the skull, causing fluid to accumulate in the spinal cord.

While it is not known how widespread SM is in Chihuahuas, multiple cases have been reported. Other affected toy breeds include Brussels Griffon, Maltese, Pomeranians and Yorkshire Terriers. Essentially any brachycephalic or toy breed can be affected or is predisposed to developing SM as shortened skulls appear to be a risk factor, says Clare Rusbridge, BVMS, DECVN, Ph.D., MRCVS, a researcher who studies the genetics and mechanisms of the

disease at the Stone Lion Veterinary Hospital in London.

SM affects individual dogs differently, with disease severity relating to the cross-sectional width of the syrinx and the area of the spinal cord that is damaged. The hallmark sign is pain, but it may occur intermittently, and owners and veterinarians inadvertently may dismiss it. Dogs don't always show signs of SM, particularly those that are less severely affected. Some may not experience pain until their senior years.

The Chihuahua Club of America (CCA) is taking steps to educate owners and breeders about syringomyelia, with a goal of reducing disease incidence through selective breeding practices. Sherri Chavez, chairwoman of the CCA Health Related Issues Committee, says, "We don't think that syringomyelia is a big problem in our breed, but it is important to work together to be sure it doesn't become one."

An MRI provides a definitive determination of SM, but the imaging is expensive. "Our goal is to encourage veterinarians to conduct a limited MRI for detection of CM and SM of the upper neck at \$500 to \$700 each, a rate we hope will encourage more people to test their Chihuahuas," Chavez says. "Once a breeder knows whether a dog is affected by the disease, he or she can make prudent breeding decisions."

A Surprising Diagnosis

A diagnosis of SM can be surprising to owners, particularly since some dogs are asymptomatic. Chihuahua breeder Virpi Kauppinen of Isalmi, Finland, was taken aback when she learned a 10-month-old Long-Coat Chihuahua out of a bitch she bred that a friend owned and bred had SM. An MRI was performed on the puppy's 3 1/2-year-old dam. Though the dam did not have signs of SM, the MRI indicated she had the disease plus CM.

Kauppinen then had the puppy's 8-year-old granddam, whom she owns, scanned. The test showed the granddam also had SM but not Chiari-like malformation. "I found myself with three

Researchers Seek SM DNA Samples

Ongoing collaborative research to identify the mutations causing syringomyelia (SM) in toy breeds continues at the Stone Lion Veterinary Hospital in London and the University of Montreal in Canada. DNA samples of affected dogs are requested as follows:

- Chihuahuas shown by MRI to have or not have SM with or without Chiari-like malformation
- Any toy breed, other than Cavalier King Charles Spaniel, shown by MRI to have or not have SM with or without Chiari-like malformation
- Any toy breed older than 5 years of age shown by MRI to have Chiari-like malformation but not SM.

Prior to submitting DNA samples, please contact Dr. Clare Rusbridge at Stone Lion Veterinary Hospital at neuro.vet@btinternet.com or 020-8946-4228 or research assistant Penny Knowler at penny.knowler@ntlworld.com. For additional information about syringomyelia, you may visit Rusbridge's website at www.veterinary-neurologist.co.uk.

Continued on page 2

Syringomyelia

continued from page 1

generations affected by SM," Kauppinen says. "The sires were from different bloodlines and had not been scanned."

The most common sign of SM is pain or irritation near the neck region, which accounts for syringomyelia being called the "neck scratcher's disease." Dogs adopt a "nose down" position when their head or neck hurts. Owners often report their dog's pain is worse at night, when waking up, during extreme temperature changes or during excitement or stress. Besides the neck, affected dogs may scratch at the shoulder, ear, chest or flanks. They typically scratch one side of the body, sometimes never making skin contact.

A complex disease, syringomyelia can occur secondary to other health conditions, such as following a spinal fracture or trauma. A tumor or other mass also can lead to obstruction of the cerebrospinal fluid pathways, resulting in SM. The most common cause of SM in toy dogs is Chiari-like malformation in which the skull may simply be too small to hold the brain.

Joyce Ferraro of Middle Grove, N.Y., first noticed her 2 ½-year-old Long-Coat Chihuahua, "Mandy," vomiting a small amount of fluid, and then the dog's tail went down and she stopped eating. A preliminary diagnosis indicated gastrointestinal problems, but blood tests and radiographs did not support the diagnosis.

As Mandy's condition worsened, she began pacing and veering into tables and chairs, unable to walk in a straight line. The dog jerked her head slightly and would hide. "We did not know she was in pain and continued to think it was a gastrointestinal problem," Ferraro says.

Ferraro took her dog to a veterinary neurologist who conducted an MRI that determined the correct diagnosis of syringomyelia. The specialist treated Mandy, prescribing prednisone and pain medication.

"Today, Mandy is about 98 percent back to normal," says Ferraro. "She plays all the time, and she is eating, jumping up on the sofa, barking and walking well."

Veterinarians manage SM medically and/or surgically, depending on the severity of the disease. Pain relief is the main objective, and non-steroid anti-inflammatory drugs, opioids or neurogenic pain killers typically are prescribed. Medications, such as prednisone, may be used in severe cases to alleviate pain and reduce cerebrospinal fluid production.

One researcher reported that one-third to one-half of dogs receiving medications eventually stopped responding to drug therapy within one and a half to three years.¹ Eventually dogs were euthanized due to unrelenting pain.

Young dogs with severe signs and pain should be considered for surgical treatment to help minimize progression

of the disease as they age. The most common surgery is an occipital decompression in which the foramen magnum, the opening at the base of the skull, is enlarged by removing a portion of the bone at the back of the skull and also part of the first neck vertebrae. This allows the passage of cerebrospinal fluid into the vertebral canal.

The procedure generally is successful in alleviating pain and improving neurological functioning. Some dogs might scratch or show discomfort years later due to the development of scar tissue and require repeat surgery.

Many dogs with SM can lead a normal life without medications or

"AN MRI SCREENING AT 5 YEARS OF AGE GIVES A TRUE STATUS OF THE DISEASE."

CLARE RUSBRIDGE, BVMS, DECVN,
PH.D., MRCVS, STONE LION VETERINARY
HOSPITAL, LONDON

surgery unless their signs worsen. Some dogs stay active and participate in sports, such as obedience or agility. Fitting a dog with a harness instead of a collar will help reduce irritation to the neck in some cases.

"The prognosis for affected dogs is hard to tell," Rusbridge says. "It is difficult to get reliable data, especially since many dogs live with SM that we don't even know about. In one study we performed, around 40 percent of dogs were eventually euthanized as a consequence of the disease."

No Simple Mode of Inheritance

In an effort to discover the gene mutations for syringomyelia, Rusbridge aims to better understand the disease to help advance diagnoses and treatment. Rusbridge believes that SM does not have a simple mode of inheritance and is likely to be influenced by other unknown factors.

Through collaborative work, Rusbridge and Zoha Kibar, Ph.D., assistant professor of the University of Montreal, have narrowed down SM candidate genomic regions in Cavaliers to two or more loci. Rusbridge believes these genes interact to cause the disease. The finding is based on genealogy information from 24 generations and more than 10,000 Cavaliers.

"We do not know whether it is the same genes in Chihuahuas as in Cavaliers or whether SM in Cavaliers with and without Chiari-like malformation is the same as occurs in Chihuahuas," Rusbridge says.

"We would like to assume it is the same disease, but we can't say for certain," Kibar adds.

Information about the relevance of SM in Cavaliers compared to other breeds is likely to provide insights. "Any research has the potential to benefit other dogs with similar diseases," says Rusbridge. "We currently are fine mapping the SM candidate loci in Cavaliers. This involves extensive genetic studies

of larger Cavalier cohorts with additional genetic markers selected from these candidate loci.

"We will next sequence all genes residing in these refined candidate regions and identify the causative mutation(s) by comparing the sequences of affected dogs to those of unaffected dogs. Finding the genes and the underlying pathogenic mutations in SM could happen in the next year but more likely in the next five years."

The long-term goal is to develop a noninvasive genetic test that can be used to identify affected dogs and carriers so breeders can selectively breed against syringomyelia. Until a test is available, breeders should have MRIs taken of potential breeding stock, Rusbridge advises.

"An MRI screening at 5 years of age gives a true status of the disease," says Rusbridge. "If a dog is clear at 5 years old, he or she is unlikely to have SM with or without Chiari-like malformation. A dog diagnosed at age 6 would have had syrinxes at 5 years of age. Popular stud dogs obviously have a great deal of genetic influence, and determining their SM status should be a priority."

According to Rusbridge, the basic principles breeders should use in determining dogs to breed are:

- At least one breeding partner — either the sire or dam — should be MRI tested to determine whether he or she is free of SM at 2 ½ years of age. Ideally, both the sire and dam are SM-free.
- If an SM-affected dog is used for breeding to preserve desirable traits or increase genetic diversity, then the chosen mate should be an SM-clear dog that is 5 years of age or older.
- In breeds like Chihuahua that have a lower incidence of CM and SM, it is best to avoid breeding dogs with CM. If CM becomes "typical" in a breed — as it is in Cavaliers — then it may be difficult to breed away from. If a dog with CM is bred, the potential mate should be determined by MRI to be CM-free and the offspring should be MRI scanned to select those without the defect for breeding.

Though understanding the genetics of syringomyelia is a slow process, research is under way to learn the mechanics of this disease and its impact on Chihuahuas and other toy breeds. Fortunately, breeders and owners take seriously the potential long-term effects of the disease and are working together to aid understanding that will benefit Chihuahuas everywhere. ■

¹ Dewey CW. *Chiari-like Malformation & Syringomyelia: A Handbook for Veterinary Professionals*. 2010;44.

Purina appreciates the support of the Chihuahua Club of America and particularly Sherri Chavez, chairwoman of the CCA Health Related Issues Committee, in helping to identify topics for the *Purina Pro Club Chihuahua Update* newsletter.