Degenerative Myelopathy

Degenerative myelopathy (also known as DM, German shepherd degenerative myelopathy, or chronic degenerative radiculomyelopathy) is a slowly progressive disease that affects the spinal cord and a dog's ability to walk. DM results in lost coordination of the hind legs, which progresses to weakness and then to paralysis of the hindquarters.

What happens is that the structures within the spinal cord that are responsible for nerve impulses degenerate. In degenerative myelopathy, the myelin (the insulation around the nerve fibers) and the nerve fibers that carry signals to the muscles do not communicate so the nerve's signals to move are not followed. While these changes can happen anywhere along the spinal cord, they usually happen in the mid to lower back.

Typically, degenerative myelopathy isn't seen in dogs under the age of five. The degeneration occurs slowly over a period of several months. Often the first signs noticed are difficulty in the hind quarters when the dog is getting up. This awkwardness is most noticeable when the dog walks on a smooth surface. However, as the disease progresses, the dog becomes uncoordinated and will scuff or drag the rear feet, causing excessive wearing of the toenails.

Sometimes one side is more noticeably uncoordinated than the other. The disease can either wax and wane episodically or progress steadily. It usually takes a few months to a year after onset for a dog to become unable to walk.

Cause

The cause is a DNA mutation in a gene called superoxide dismutase 1 (SOD1). This risk factor of having this gene was identified in 2009. Prior to this discovery, genetic, nutritional, and immune factors were suggested as possible causes of DM.

The neurologic disease is similar to some forms of human amyotrophic lateral sclerosis (ALS, often called Lou Gehrig's disease) and DM usually affects dogs that are between 8 and 14 years old. Though most of the dogs in early reports were German Shepherd Dogs (GSD), other breeds that have the mutation and clinical signs include American Water Spaniel, Bernese mountain dog, Bloodhound, Borzoi, Boxer, Canaan Dog, Chesapeake Bay retriever, English Cocker Spaniel, German Shepherd Dog, Great Pyrenees, Kerry blue terrier, Pembroke Welsh Corgi, Pug, Sealyham terrier, and Whippet.

Symptoms

Early signs, such as difficulty getting up or a noticeable sway in the dog's gait, may be confused with hip dysplasia. As months go by, "scuffing" of the hind limb toe nails or dragging of the hind limb feet usually is noted. The rear limbs may criss-cross when standing or walking. The dog may not be able to stand well and during an examination a veterinarian will detect rear limb weakness. Muscles of the rear limbs will become atrophied or wasted away. As the disease progresses, the front legs may become weak. Fortunately, dogs with DM do not appear to be in pain.

Diagnosis

Since the clinical signs of DM can appear the same as those for other neurologic diseases such hypothyroid neuropathy, protozoal infections and other problems, the diagnosis of DM is based on excluding these diseases. Thus a variety of diagnostic tests are needed, including physical and neurological examinations, routine blood work, spinal x-rays, genetic testing and spinal tap. Sometimes referral to a specialist is required, for advanced imaging such as myelogram, computed tomography (CT scan) or magnetic resonance imaging (MRI) scans. Myelogram is a study where die is injected into the fluid around the spinal cord to look for compression. With DM, that study will be normal, and there will be no compression. Unfortunately, it is possible that performing myelogram on a dog with DM can make things worse. In the end, a definitive diagnosis can only be ascertained after death by looking at biopsy samples from the spinal cord.

DNA testing by the Orthopedic Foundation for Animals and many other services can identify the DNA mutation that is associated with the development of DM. The test identifies dogs that are clear and have two normal copies of the gene, carriers who have one normal copy and one mutated copy, and those who are at much higher risk for developing DM because they have two mutated copies. In unpublished studies, dogs who have two of the abnormal genes are the only ones that have developed the disease. However, there are some dogs in these studies with two mutated genes and without clinical signs; it is unclear yet whether these dogs will later develop the disease or if other factors are necessary for DM to develop.

The DM DNA Test may be performed on any breed or mixed (hybrid) breed. However, the test is sometimes not accurate in the Bernese Mountain dog, which can have a false positive. By testing before breeding, breeders will be able to avoid mating two dogs with the mutated DNA. As part of an ongoing collaborative effort by research scientists at the University of Missouri and the Broad Institute, a DNA test is available for dogs that have been diagnosed with DM, and for older dogs in selected breeds. Details are outlined in the Research section of that website.

Treatment

Unfortunately no treatment has been shown to reverse the signs; DM can be managed but not cured. Supportive treatment can help.

1. Exercise such as walking and swimming should be encouraged. Physical therapy helps to maintain muscle mass and quality of life. In one study of 50 dogs with DM, those that received intensive physiotherapy had longer survival time (mean of 255 days), compared with dogs that received moderate (mean of 130 days) or none (mean of 55 days). Affected dogs who received physiotherapy were ambulatory longer than those that did not receive it.

2. Once the dog reaches the non-ambulatory state, pressure sores, urine leaking, and loss of bowel control are likely to develop, so close attention to daily eliminations and bedding will be needed.

3. A combination of epsilon-aminocaproic acid, N-acetylcysteine, prednisone, vitamins B, C, and E, and exercise therapy has been suggested to slow the progression. However, this regime may have no benefit over doing only the physical therapy. A study evaluating combination therapy with aminocaproic acid, N-acetyl- cysteine with vitamins B, C, and E failed to detect a benefit.

Breeding and Prevention

Because the disease is found in specific breeds, responsible breeding is the only way to prevent degenerative myelopathy. If you plan to get a purebred puppy of an affected breed, ask the breeder about history of DM in the kennel's line. Understand that clinical signs don't develop until long after sexual maturity.

The DM gene mutation is very common in some breeds, such as the German shepherd. Overly aggressive breeding programs to remove the gene mutation may create a bottleneck effect, possibly increasing the risk of other diseases and eliminating other desirable qualities of the breed. A realistic approach is to consider the DNA DM results as you would consider any other undesirable trait or fault. Dogs testing at risk should be considered to have a more serious fault than those testing as carriers.

Prognosis

Most affected dogs are euthanized due to disability within 6 to 12 months of onset of signs.

References:

Becky Lundgren, DVM – VeterinaryPartner.com
Kathmann I et al. Daily controlled physiotherapy increases survival time in dogs with suspected degenerative myelopathy. *J Vet Intern Med*. 2006 Jul-Aug;20(4):927-32.
Wendy Blount, DVM – PracticalVetMed.com