

# Degenerative Myelopathy

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## Background

Degenerative myelopathy (DM) is a progressive canine spinal cord disease characterized by axon and myelin degeneration. The white matter tracks of the thoracolumbar spinal cord are most severely affected. As the disease progresses, lesions will also appear in the lumbosacral and cervical spinal cord. Clinical signs follow this progression. DM is purely a degenerative process, there is no inflammatory component.

## Presentation

Signalment - Most dogs with DM present at the age of 8 years or older (occasionally dogs can present younger). Commonly affected breeds include the German Shepherd dog, Pembroke and Cardigan Welsh Corgi, Boxer, Chesapeake Bay Retriever, and Rhodesian Ridgeback; however the disease has been seen and confirmed in numerous other breeds, as well as mixed breed dogs.

History – Dogs with DM usually have a chronic, progressive history of pelvic limb weakness and incoordination. Owners may describe that their dog has more difficulty getting up, walking on slick surfaces, going up and down stairs, and they may also describe dragging or knuckling of the back feet. Urinary and/or fecal incontinence may also be reported.

Neurologic examination findings – When patients are examined early on in the course of disease, ambulatory paraparesis with pelvic limb ataxia is the most common finding. Proprioception in the pelvic limbs is decreased to absent and asymmetry is common. Over time this progresses to paraplegia and eventually tetraplegia. Spinal reflexes will be intact in the earlier stages, reflecting the lesions of the T3-L3 spinal cord. As the disease progresses and the lower motor neurons of the lumbosacral and eventually the brachial plexuses become affected, the spinal reflexes may be decreased to absent. With time, urinary and fecal incontinence will develop, along with a flaccid bladder and decreased anal tone. In its final stages the muscles of respiration can lose their innervations as well, however most dogs are euthanized long before this occurs. DM is a nonpainful disease, so spinal palpation should not elicit a reaction. It is important to keep in mind that conditions such as lumbosacral disease and coxofemoral osteoarthritis are also common in older, large breed dogs and their concurrent presence can confound this aspect of the history and/or examination.

## Diagnosis

The only way to obtain a definitive diagnosis of DM is with histopathologic examination of the spinal cord at postmortem. During life, we can achieve a presumptive diagnosis of DM by exclusion of other myelopathies. Following a complete neurologic examination and general health evaluation as indicated for each patient, advanced diagnostic including an MRI (ideal) or myelogram, and cerebrospinal fluid (CSF) analysis should be performed. The advanced imaging will help to rule out other nonpainful myelopathies including neoplasia and myelitis. Compressive myelopathies such as intervertebral disc disease, extradural neoplasia, and intradural-extramedullary neoplasia can be ruled out as well. The cerebrospinal fluid will help rule out inflammatory myelopathies. In dogs with DM, the results of the advanced imaging and CSF analysis should be normal. It is not uncommon to see evidence of intervertebral disc degeneration with varying degrees of extrusion or protrusion; however the degree of epidural fat and cerebrospinal fluid attenuation can be evaluated to determine the likely clinical significance of these lesions. A lack of spinal pain will also support the fact that these lesions may be clinically insignificant. Electromyography and nerve conduction studies are also indicated to help with diagnosis in some cases.

## New Developments in Diagnosis

Thanks to combined efforts by the University of Missouri and the Broad Institute of MIT/Harvard, a mutation in the superoxide dismutase 1 (SOD-1) gene has been discovered in dogs affected with DM. This same gene is mutated in human patients with familial amyotrophic lateral sclerosis (ALS), or Lou Gehrig's disease. A test for the presence of this mutated gene is now available and being offered through the OFA. The test can be done either on an EDTA blood sample or a cheek swab. Care must be taken to ensure that the owners are counseled by a veterinarian regarding the results in light of that particular dog. The counseling veterinarian should not only understand the test, but also should be familiar with the patient and its clinical signs. The test will reveal if the patient is homozygous for the normal SOD-1 gene, homozygous for the mutated SOD-1 gene, or heterozygous.

**Homozygous normal gene (clear)** – It is highly unlikely that a patient's signs are due to DM if this is the test result. It is also unlikely that this dog will develop DM in the future.

**Heterozygous (carrier)** – It is still unlikely that this patient's signs are due to DM and unlikely that they will develop DM in the future.

**Homozygous affected/mutated gene (affected)** – If the clinical signs are consistent with those of DM, and if the advanced imaging and CSF analysis are normal, it is likely (but not definitive) that this dog has DM. It is also possible that a dog with this test result could develop DM in the future. At this point, all dogs that have been studied with a confirmed diagnosis of DM have had this test result. However, numerous dogs with this test result have not had, nor have they developed DM. Therefore, the terminology of “affected” that is used for this category can be misleading when interpreted by an individual unfamiliar with the test.

The DNA test can be used to aid in a presumptive diagnosis in a dog with clinical signs; however it is also being used for screening by many breeders. At this time researchers are not recommending removal of dogs with carrier or affected test results from the breeding population. Rather, they are encouraging breeders to consider a test result as one factor when planning an optimally balanced breeding. Since this test is still relatively new, the total number of dogs with a DNA test and postmortem examination of the spinal cord is not high and this should be considered when interpreting test results. Presence of the mutation is clearly a risk factor in development of disease; however other, currently undefined, risk factors also clearly exist. This test provides a helpful piece of information, but histopathologic evaluation of the spinal cord is still the only way to reach a definitive diagnosis of DM. Researchers are aggressively pursuing this disease in many breeds to rapidly increase this pool of data. Consequently some dogs may be eligible for free DNA testing. The Canine Genetic Disease Network is an excellent resource for both veterinarians and clients: it provides a summary of the disease, a link to the OFA website for sample submission, guidelines on which dogs are eligible for free testing, and information for breeders. The website is <http://www.caninegeneticdiseases.net/DM/mainDM.htm>

### **Treatment and Prognosis**

Unfortunately there is no treatment for this progressive disease. Most dogs will progress to nonambulatory status within 6 months following a presumptive diagnosis and most will be euthanized within 6 months to 1 year. A study looking at the disease in the Pembroke Welsh Corgi did show an exception to this, with the clinical course extending as long as 19 months after a presumptive diagnosis. This may be due to owners being more willing and able to manage nonambulatory dogs that are smaller in size. While various vitamins, supplements, and other nutraceuticals, including amino-caproic acid, have been touted by various sources, no studies have ever been published to support the benefit of any of these. Many of these medications are quite costly and can cause gastrointestinal upset, so their use is not recommended. Since there is no inflammatory component to this disease, steroids are not indicated.

One study did show that dogs with DM who received regular, controlled exercise had a longer clinical course before euthanasia than those that did not. While many of these dogs did not have a definitive diagnosis, controlled activity, exercise, and rehabilitation are inexpensive, without side effects, and also helpful for many of the other diseases that can be top differentials for dogs with DM. They also provide the client with an opportunity to be an active part of their pet's treatment, enhancing their bond. Formal consultation with a canine rehabilitation practitioner is ideal so that a comprehensive plan can be made for activities that can be performed within a rehabilitation practice and/or at home.

Supportive and nursing care is a crucial component to optimal treatment of dogs with DM. Patients often benefit from booties to protect the dorsal pedal surface during the stages where the patient is still ambulatory to prevent sores and abrasions. Keeping the patient clean, dry, and providing ample padded bedding is important to prevent decubital ulcers. Any sores that do arise should be treated immediately and aggressively, as they can progress quite rapidly to severe, infected ulcerations. As the patient becomes weaker, slings and wheelchairs can be used to improve patient mobility. Finally, bladder management is imperative when the patient progresses to the point of incontinence. Urinalysis and culture should be performed regularly (and whenever clinical signs of urinary tract infection arise) so that infections can be treated in a timely manner.

### **Client education**

Obviously it is important to counsel owners regarding the progressive nature of the disease. Every owner has different limitations for how much supportive care they are willing and able to provide. They also have variable outlooks on the level of function that their pet much have for a given perceived quality of life. Discussions about the appropriate time for euthanasia must take owner limitations as well as patient condition into account. Before the time comes for these difficult decisions, owners should be educated as to the nursing care issues mentioned above. They should also be told what to watch for that may indicate important changes in their animal's status, such as loss of urinary continence. When this arises, the owner should learn appropriate bladder management techniques. Recognizing changes in urine color and odor can help identify urinary tract infections so they can be treated early in their course. The Canine Genetic Diseases website provides multiple helpful, reliable links for clients to explore.

### **Summary**

Degenerative myelopathy is a progressive, nonpainful spinal cord disease that affects older dogs. Lesions are of axonal loss, not inflammation, and begin in the

T3-L3 spinal cord segments. With time, lesions will also appear in the lumbosacral and cervical spinal cord segments as well. Clinical signs progress along with these localizations. Most dogs will progress to the point where the client elects euthanasia by 6 months to 1 year following the onset of clinical signs; however this tends to be extended in smaller dogs. A definitive diagnosis of DM cannot be achieved antemortem, but advanced spinal cord imaging and CSF analysis are used to rule out other potential etiologies. There is now a DNA test available to detect the genetic mutation that is a risk factor for disease, but these results must be interpreted in light of each individual case. While there is no cure for DM, controlled exercise, proper nursing care, and client education can optimize the dog's quality of life, and possibly even its longevity.