# Meningoencephalitis of Unknown Origin

### **ABOUT THE DIAGNOSIS**

Meningoencephalitis of unknown origin (MUO) is a small group of inflammatory disorders of the brain, spinal cord, and/or optic nerve. These disorders include granulomatous meningoencephalomyelitis (GME), necrotizing leukoencephalitis (NLE), and necrotizing meningoencephalitis (NME). These specific disorders can only be differentiated from one another based on brain biopsy, or on a postmortem exam. Because brain biopsy is difficult and risky, it is seldom performed. Instead, the diagnosis centers on "ruling out" other causes of central nervous system (CNS) disease. The fundamental problem is inflammation (-itis) of the lining of the brain and spinal cord (meninges) and the brain (encephalo) and spinal cord (myel). The cause of MUO is unknown, but misdirected response of the body to an infection (where neurologic tissue is damaged as an "innocent bystander") or an inadvertent, self-targeted attack of the CNS by the body's own immune system are suspected. Regardless of specific trigger, the damage is done by the body's own system of inflammation being overly active in and around the brain, which is harmful.

Dogs with MUO are most often between 1 and 8 years of age, and small breed dogs and Airedale terriers are more likely to be affected than other dogs. These inflammatory disorders occur in one of two forms: focal, where the symptoms can be localized to one area of the brain or spinal cord, or multifocal/disseminated, where multiple areas of the nervous system are affected.

This disease is confined to the central nervous system (brain and spinal cord), with the nerves to the eye (optic disc) affected on occasion. A variety of symptoms can occur, depending on the region of the brain or spinal cord involved. Possible symptoms include seizures, blindness, walking aimlessly in circles, holding the head in a sideways-tilted position, weakness or paralysis of the legs, reluctance to move, and neck pain. These symptoms appear suddenly, although in the focal form, the disease often advances more slowly, and symptoms may appear more gradually, over a period of days or weeks, compared to the disseminated form.

There are many other possible causes of these types of symptoms, and adequate medical treatment depends on confirming whether MUO, or an altogether different type of neurologic problem, is present. Basic information that helps your veterinarian assess the likelihood of MUO versus other, "impostor" syndromes that mimic MUO include: information from you regarding duration and type of symptoms you or others have observed; physical and neurologic examination performed by the veterinarian; routine blood and urine tests; and routine x-rays and ultrasound exams of the chest and abdomen in some cases. There are two primary diagnostic tests for determining whether MUO is present: imaging studies and examination of cerebrospinal fluid (CSF). Imaging studies, such as MRI or CT scans, are used for looking at the structure of the brain and are especially useful in cases of focal disease. These advanced imaging studies require general anesthesia and special equipment; they are typically performed by a veterinary neurology specialist. For CSF testing, the pet is under general anesthesia to prevent movement and to avoid pain. A spinal tap is performed, during which a small needle is inserted into the space around the spinal cord, either at the base of the skull or at the lower back. A small amount of spinal fluid is withdrawn and submitted for microscopic examination and laboratory testing. The spinal fluid of dogs with MUO usually has characteristic abnormalities that set it apart from other types of central nervous system diseases. To eliminate the possibility of infectious disease as a cause of the symptoms, since both the symptoms and the spinal fluid from severe infections can be similar to those for MUO, specialized blood screens for certain infections and spinal fluid bacterial or fungal cultures may be necessary. Such extensive testing is typically necessary because MUO is an incurable, progressive disease. It can be treated with medications to improve/reduce symptoms, but it is preferable to find another, curable problem as the cause of symptoms instead, and finding a different disease that imitates MUO but can be permanently eliminated is one of the main purposes of the diagnostic medical tests listed above.

## LIVING WITH THE DIAGNOSIS

The range of response to medications and outcome is extremely broad: some dogs respond to treatment and survive for months, or even years, while others die within days to weeks, despite treatment. One of the most important determinants of how a dog will do is his/her response to initial treatment after the diagnosis of MUO has been confirmed with a spinal tap.

#### **TREATMENT**

Since the fundamental problem is a misdirected immune attack on the brain tissue, immunosuppressive medications help some dogs. Corticosteroids (cortisone-like medications) are given to try to reduce the degree of immune-mediated damage. You can give these medications as tablets by mouth at home. In dogs that respond favorably, treatment is lifelong. Discontinuation of treatment can result in a relapse of the disease, so be sure to give the medication consistently and if you must stop for some reason, contact your veterinarian first to discuss an alternative. In many instances, other chemotherapy or immunosuppressive drugs can be tried. These have the advantage of greater effectiveness when used properly, and fewer cortisone-associated side effects. They should be used in consultation with a veterinary neurologist (see fourth item in "DOs," below). In some cases, radiation therapy may be effective for dogs with the focal form of MUO. Severely affected or unstable pets may need to be hospitalized for initial treatment, and the degree to which any pet responds to treatment is highly variable and cannot be predicted.

#### DOs

- Realize that MUO is a serious but potentially manageable disease that almost always requires lifelong treatment. Very roughly, one third of affected dogs do well long term, one third die very soon after diagnosis, and one third can survive with a good quality of life for some months.
- Be attentive to the specific needs of a pet with MUO: vision loss and possible disorientation mean that certain situations should be avoided. Examples of activities or circumstances that a pet with MUO should avoid include: walking beside steep drops (long flight of stairs, edge of a cliff), swimming in deep water, and any other situation where decreased vision or impaired mental function could bring harm to a dog that is compromised.
- Give the medication as directed, and if you notice an increase in panting, restlessness, and excessive drinking and urinating, realize that these may be medication-related, not pain or discomfort.
  If such symptoms appear excessive, you should notify your veterinarian, and an alternative treatment approach may be preferable.
- Consider seeing a veterinarian specialized in neurologic disorders if the diagnosis of MUO is uncertain, or for the latest treatments, or

simply for a second opinion. Veterinary neurologists are known as Diplomates of the American College of Veterinary Internal Medicine or the European College of Veterinary Neurology and directories are available at www.acvim.org or www.vetspecialists.com and www.ecvn.org.

Sheets:

Other information that may be useful: "How-To" Client Education

- How to Change the Environment for a Pet That Is Blind
- How to Manage a Pet That Is Having Seizures

#### DON'Ts

• Do not discontinue corticosteroid treatment abruptly. This is likely to cause a relapse.

## WHEN TO CALL YOUR VETERINARIAN

• If your pet's symptoms worsen.

# SIGNS TO WATCH FOR

Worsening of symptoms most commonly emerges as one or more of the following:

- · Seizures.
- Weakness or paralysis of the limbs.
- · Neck pain or reluctance to move.
- Blindness.
- · Circling, head tilt, or pressing the head against walls.

# **ROUTINE FOLLOW-UP**

 Dogs on immunosuppressive therapy will need periodic examinations and laboratory testing to detect possible side effects of treatment and allow the treatment to be adjusted as needed.

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