

Lymphangiectasia

ABOUT THE DIAGNOSIS

Cause: Lymphangiectasia is a protein-losing intestinal disease of adult dogs. With lymphangiectasia, a disruption of the lymphatic system causes leakage of protein-rich lymphatic fluid (also called lymph) into the gastrointestinal tract. This loss of lymph through the feces (stool, excrement) means the proteins within it leave the body and cannot be used for building and maintaining tissues, muscle bulk, and strength.

The lymphatic system is a network of fluid, vessels, lymph nodes, and organs throughout the body that has numerous functions. It often runs parallel and adjacent to the blood circulation. The lymphatic system is a ferrying system that carries waste substances outward from body tissues to the bloodstream. It also provides immune defense in certain areas of the body such as the spleen, tonsils, and the lining of the gastrointestinal tract (stomach and intestines). Also in the intestine, the lymphatic system absorbs fats after they are digested (chyle). In addition to fats, lymphatic fluid contains proteins and white blood cells, which are vital for the body's functions. Unfortunately, with lymphangiectasia the lymphatic circulation is disrupted and white blood cells, proteins, and fats leak into the intestinal tract and are wasted. As a result, the dog becomes malnourished. Over time, this potentially can become a life-threatening disease.

Primary lymphangiectasia is thought to be present at birth (congenital); however, symptoms are usually seen later. Although the intestinal lymphatic system is usually affected, other signs include the accumulation of a milky-looking, chylous effusion around the lungs (chylothorax), edema or swelling under the skin precipitated by decreased protein in the blood (subcutaneous edema), and fluid in the abdominal cavity (ascites).

Secondary lymphangiectasia has many potential causes. These include inflammation of the intestine, heart problems that cause right-sided congestive heart failure, obstruction of the thoracic duct (the thin vessel that carries lymphatic fluid from the abdomen and part of the chest to the bloodstream), and certain types of intestinal cancer.

The exact cause of lymphangiectasia often cannot be determined despite extensive testing, and a large proportion of dogs with lymphangiectasia do not have any of the disorders listed above (no inciting cause is ever found).

Although soft-coated wheaten terriers, Yorkshire terriers, and Norwegian lundehunds are most commonly affected with lymphangiectasia, any breed of dog can be affected. This disorder is very uncommon in cats.

Besides weight loss, there are other common symptoms in dogs with severe lymphangiectasia. They often develop a big belly, despite a loss of muscle and fat, due to fluid accumulation. The rib cage and spine may be easily seen and felt although the belly looks swollen. Dogs with this disorder are prone to develop blood clots, either in the lungs or the large blood vessels in the abdomen. Either blood clots or abdominal effusion (fluid accumulation) can cause difficulty breathing. Diarrhea, and sometimes vomiting, might be seen in dogs with lymphangiectasia. These dogs often have a poor appetite.

Diagnosis: When lymphangiectasia is suspected, a complete blood count (CBC), serum biochemistry profile, urinalysis, and fecal analysis are performed to look for characteristic changes associated with this disease, to assess overall health, and to rule out other possible

causes that could be mimicking lymphangiectasia. X-rays of the chest and abdomen may be taken to screen for fluid accumulation or signs of any inciting causes. A fine-needle aspirate helps to characterize the type of effusion in the chest and/or abdomen when present. For this procedure, a very small needle is inserted into the body cavity without the need for anesthesia; fluid is aspirated and examined under a microscope. Lymphangiectasia is ultimately diagnosed from a biopsy of the gastrointestinal tract that is obtained either endoscopically or during a surgical procedure. That is to say that lymphangiectasia can only be suspected, but not pinpointed, until a sample of intestinal tissue is examined by a pathologist to confirm lymphangiectasia and rule out all other possible intestinal diseases that produce similar or identical features. The biopsied intestinal tissue is submitted to a laboratory where a specialist examines it under a microscope to make the diagnosis; therefore, it is common for a period of 3-5 days to elapse after the biopsy procedure before the lab's diagnosis is known.

LIVING WITH THE DIAGNOSIS

Except for the unusual cases where a curable cause for lymphangiectasia is found (that is, secondary lymphangiectasia), most dogs will need to live with the disorder for life. Treatment can be challenging, but many dogs are well managed for long periods of time.

The cornerstone of treatment is your dog's food. It is quite a challenge to get just the right balance of nutrients into these patients; typically, a very-low-fat diet is necessary but these very thin dogs need lots of vitamins and just the right balance of protein and carbohydrates. There are prescription diets available only through a veterinarian that can serve this purpose for most dogs. If you prefer, you may make your dog's diet yourself, although it is critical to offer the correct balance of nutrients for dogs. You should seek the recommendations of a veterinary nutritionist (Diplomate of the American College of Veterinary Nutrition; directory at www.acvn.org) because there are many, many impressive-sounding diets on the market, in books, or on the Internet, but most do not meet the unique needs of a dog with lymphangiectasia.

TREATMENT

A low-fat and highly digestible diet that is calorie dense is an important part of therapy, along with supportive and specific care. If an underlying disease can be identified, it must be treated. Because a cause for lymphangiectasia is usually not determined, the symptomatic and dietary treatments are usually required for life. In addition to the strict dietary restrictions, an antiinflammatory medication (corticosteroid or cortisone-derivative) may be given. If enough fluid has accumulated in the abdomen (belly) to make breathing difficult, this may need to be drained. Usually, drugs are given help to prevent blood clots from forming abnormally. Most dogs will be given B vitamin supplements as well.

DOs

- Inform your veterinarian if your cat or dog has ever been diagnosed with a medical condition and is taking medication, because this information may increase or decrease the importance of performing certain tests, influence which medications should be used for the lymphangiectasia, and may even affect the prognosis (outlook for long-term recovery).
- Give medication exactly as directed by your veterinarian, and if you are concerned about possible negative effects, discuss them

with your veterinarian immediately rather than simply discontinuing the treatment.

- Realize that getting to the point of having a conclusive diagnosis of lymphangiectasia is always challenging because of the tests and biopsies required to reach the diagnosis. Furthermore, the diagnosis can be disappointing since there is no cure. However, individual response to treatment is highly variable, and often the only way to know if treatment will work for a particular dog, once the diagnosis of lymphangiectasia is made, is to try treatment and monitor response. Some dogs do very well while others do poorly, and often the only way to know for sure is to try.
- Consider seeking consultation with veterinary specialists trained to diagnose and treat this complicated, often frustrating disease. Specialists of the American (or European) College of Veterinary Internal Medicine are found in most larger cities in North America and Europe. Your veterinarian can offer a referral, or you can find them at www.acvim.org, www.vetspecialists.com, or www.ecvim-ca.org.

DON'Ts

- Do not substitute another diet without consulting your veterinarian first. Malnutrition is a common, life-threatening complication of this disease.
- Do not feed treats that have not been approved by your veterinarian.
- Do not breed a dog that has been diagnosed with lymphangiectasia.

WHEN TO CALL YOUR VETERINARIAN

- If you cannot keep a scheduled appointment.
- If you are unable to give medication as directed.
- If your dog's symptoms are not improving after treatment begins.
- If your dog's appetite is decreasing.
- If your dog has any noticeable trouble breathing.

SIGNS TO WATCH FOR

- General signs of illness: lethargy, weakness, decreased appetite, other behavior changes.
- Signs of lymphangiectasia beginning or recurring: profound weight loss despite a strong or even ravenous appetite (polyphagia), respiratory problems (if pleural effusion is present), vomiting, diarrhea, and/or lethargy.

ROUTINE FOLLOW-UP

- Follow-up visits will be scheduled as needed (depending on the exact features of your dog's case) to monitor progress, especially involving measuring body weight and blood levels of protein and white blood cells.

Practice Stamp or Name & Address

Also available in Spanish.