

Copper Storage Hepatopathy in Dogs

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BASIC INFORMATION

Description

An important function of the liver is to remove toxic substances, such as copper, from the bloodstream. In some dogs, abnormalities can occur in the proteins that bind copper in liver cells. The liver of these dogs can still remove copper from the bloodstream, but it gradually accumulates to toxic amounts inside the liver cells. The affected cells die, which reduces the overall function of the liver. Breeds at high risk for copper storage hepatopathy include the Bedlington terrier, Doberman pinscher, Labrador retriever, cocker spaniels, West Highland white terrier, and Skye terrier. Abnormal copper accumulation can also occur with long-standing diseases of the bile system; however, the amount of copper that accumulates is significantly less than with primary copper storage hepatopathy.

Causes

In the Bedlington terrier, this disease arises from a mutation in the gene *COMMD1*, which contains the instructions for making a protein that is necessary for the excretion of copper from liver cells. In the other mentioned breeds, there is evidence that a genetic component may be involved, but the actual gene is not known at this time. Regardless of cause, clinical signs in affected dogs develop following the death of liver cells from excess copper.

Clinical Signs

Clinical signs of copper hepatopathy are the same as for most other liver diseases, and they can be vague and variable in the early phases of the disease. Dogs are often lethargic, may appear depressed, have decreased or complete loss of appetite, and have reduced exercise tolerance. In severely affected animals with diminished liver function, stupor or coma may develop, dogs may become extremely sleepy after eating, and excess fluid accumulation (ascites) may develop in the abdomen or under the skin.

Diagnostic Tests

Typically, the diagnosis is suspected when a dog of a breed known to be prone to this condition shows clinical signs of liver disease and routine laboratory tests indicate abnormal liver function. Additional laboratory tests, abdominal x-rays, and an ultrasound may be needed to confirm the presence of liver disease and to rule out diseases that can cause similar clinical signs.

Definitive diagnosis of copper storage hepatopathy requires a liver biopsy to measure the amount of copper that is present. Several methods are available for obtaining the liver biopsy, including needle biopsy under ultrasound guidance, laparoscopy surgery through a tiny keyhole incision, and a more routine surgical approach through an abdominal incision. The method chosen depends on the equipment available and the surgical experience and preference of your veterinarian. In some cases, your dog may be referred to a veterinary specialist for the procedure.

TREATMENT AND FOLLOW-UP

Treatment Options

The most important treatment for this hepatopathy is removal of the copper from the liver, which is done through a process called *chelation*. Chelation drug therapy with D-penicillamine binds the copper and releases it into the urine and feces. Once the amount of copper in the liver is decreased, steps are taken to reduce the accumulation of new copper by feeding a special diet that is extremely low in copper.

Additional drugs and supplements are commonly given, such as large amounts of vitamin C and zinc. Together, the vitamin C and the zinc make it more difficult for copper to be absorbed from the gut into the body. To work effectively, the zinc *must* be given at least 2 hours before the dog is fed. Some dogs have nausea and vomiting when zinc therapy is started, but these usually resolve within a week.

Follow-up Care

Regular follow-up visits and monitoring tests are needed during the initial phase of the therapy. A follow-up liver biopsy is often recommended after 2-3 months to measure the amount of copper present and determine whether drug therapy has been adequate. Once the low-copper diet has been started, liver tests may be rechecked every 3-6 months. Treatment and periodic monitoring are usually needed for the rest of the dog's life.

Prognosis

If the disease is caught early and treated appropriately, many affected dogs can lead normal lives. Prognosis is more guarded (uncertain) for dogs that have severe clinical signs, because they may not be able to regenerate enough liver tissue to function normally.