

Copper Storage Disease

By Dr. Karen Becker

Hi, I'm Dr. Becker. Copper is an important trace element vital to the production of enzymes and other important cellular processes. Copper in your pet's diet is metabolized and stored in the liver. The excess amount of copper is excreted in the bile and then leaves the body via stool and urine. Some animals are born with or develop changes in the liver that actually trap copper inside individual liver cells, resulting in toxicity.

Copper storage disease is exactly what it sounds like. It's an abnormal accumulation of copper in the liver, which can lead to progressive damage and scarring of the liver, which is called cirrhosis. The condition is also known as copper storage hepatotoxicosis.

Copper storage disease is most often seen in the Bedlington, Skye and West Highland White terrier, as well as the Doberman pinscher. Bedlington terriers typically acquire chronic hepatitis as the result of an inherited trait that causes copper to stay in the liver instead of being eliminated from the body. Bedlington terriers with the gene typically develop acute liver necrosis before they are 6 years of age. However, breeding management has really helped to decrease the number of cases.

In Dobermans, blood work can be abnormal as early as a year of age, but the liver disease doesn't usually become symptomatic until Dobermans are much older, around 7 years of age. In Westies and Skye terriers, the disease can occur at any time. The copper accumulation can be apparent as early as 1 year of age. Dalmatians and Labs with the condition are typically middle-aged and develop symptoms that are associated with chronic hepatitis. The condition can occur in cats, but thankfully, it's quite rare.

Factors for the Development of Copper Storage Disease

There are three factors that play into an animal acquiring this disease. Number one, failure to excrete copper in the bile. This is a common problem, more so in the Bedlington terriers. Excessive copper storage is the second reason. This means that the liver sometimes stores too much copper, especially in the cases of liver inflammation or chronic liver disease. Excessive copper ingestion or too much copper consumed at one time can overwhelm the liver's storage capacity and can cause liver damage, especially in dogs with a genetic tendency to store copper.

Copper storage disease generally falls into one of three categories: subclinical disease, which means the animal has the condition but has no obvious symptoms; acute disease, which is most often seen in young dogs and causes hepatic necrosis or death of liver tissue; and chronic, progressive disease, which usually occurs in middle-aged and older dogs existing with chronic liver disease and the condition of cirrhosis, or scarring of the liver.

Symptoms of Copper Storage Disease

Symptoms of copper storage disease depend on the amount of damage to the liver. [It] can include lethargy, anorexia, depression, vomiting, diarrhea, excess thirst, as well as urination, jaundice, which is animals turning yellow or having a yellow skin appearance; anemia, fluid buildup in the abdomen, which is called ascites; dark-colored urine and hemoglobin in the urine, which changes urine color. There can also be spontaneous bleeding and black or tarry stools from bleeding through the gastrointestinal (GI) tract.

On occasion, there can also be nervous system disorders resulting from the liver's inability to break down ammonia in the body. Your pet can actually have some neurologic stumbling and some circling conditions in advanced situations.

Diagnosis and Treatment

Your veterinarian will absolutely take a complete history of your dog and perform a thorough physical exam, as well as some diagnostic tests, which include a complete blood count called a CBC, a biochemical profile, a urinalysis and a serum bile acids test. An abdominal ultrasound will be also be performed to evaluate the condition of the liver. However, a definitive diagnosis of copper storage disease requires a liver biopsy, which will include quantifying the amount of copper that's actually in the liver. Pets with copper storage disease require lifetime treatment. As with any disorder, the earlier that you can diagnose this disorder and, of course, institute treatment, the better your dog's or cat's chances of living normally with a great quality of life.

Treatment of this condition requires removing toxic levels of copper from the body, and preventing further accumulation from occurring. The traditional veterinary approach to removing excess copper in the liver is a drug called penicillamine, which actually helps to chelate or bind it up, and then releases it from liver tissues, and promotes excretion through the urine. However, the use of copper-chelating agents in Dobermans is controversial because the disease tends to progress even when you utilize these medications.

Recommendations for Copper Storage Disease

I strongly recommend that a homemade, copper-free diet is instituted as quickly as possible. The reason I say copper-free is you want a homemade, nutritionally balanced diet to be fed to your pet. All of the nutrients need to be optimally nourished, but not copper. The diet needs to be put together by a veterinarian-nutritionist, or someone who is capable of formulating a beautifully balanced diet, minus the copper.

It's very important that these patients received optimal levels of zinc, because research shows that zinc has a really nice beneficial effect at helping to bind up copper and push it out of the system.

Research also shows that some of these pets could have problems metabolizing some of the synthetic forms of copper used in many of the bulk pet food premixes. Increased levels of zinc can actually help to reduce the amount of copper absorption in your pet's body. It's also important that the diet is formulated with nice levels of vitamin B6 or pyridoxine, because B6 can also be beneficial at helping to excrete additional copper from the body.

If pets stay on commercial diets, it's really important that they are formulated with very low levels of copper. Homemade diets need to also be sourced from low-copper sources of meat, which includes beef, white meat chicken, turkey and eggs. All of those protein sources are low in copper. It's important that organ meats be avoided because organ meats typically have higher amounts of copper.

Commercially available diets need to have a copper level that's less than 0.5 parts per million of copper. Less than 1 part per million of copper should be consumed through drinking water. If you're on well water, it's important that your well water does not contain additional copper. Vitamin C should also be avoided because vitamin C increases copper absorption. Supplements like vitamin E are recommended as antioxidant therapy, because we know increased vitamin E can help reduce the amount of liver damage that's occurring.

Additional supplements that can be very beneficial are S-Adenosyl methionine (SAME), ursodiol, as well as phosphatidylcholine, which is an anti-fibrotic agent, as well as milk thistle, which can be really beneficial at protecting healthy liver cells.

Full recovery from copper storage disease is actually quite rare, but management is usually possible. At a minimum, most dogs with the condition must be fed a very low copper diet for the remainder of their lives. This is also the very best approach for young dogs with the condition, to prevent the accumulation of copper in the liver before it becomes a major problem.

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