LIVER SHUNT
PORTOSYSTEMIC SHUNT

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Introduction

Portosystemic shunts are a common topic of conversation amongst breeders and owners of small and toy breed dogs. Congenital portosystemic shunts are being diagnosed with increasing frequency, and many breeders and veterinarians are questioning whether heredity may play a role in the spread of this disease.

What are Portosystemic Shunts?

A portosystemic shunt is a blood vessel that bypasses liver tissue, carrying blood from the intestines, stomach, spleen, and pancreas to the heart before it can be filtered and cleansed of proteins, sugars, bacteria, and toxins. Shunts are present in all fetal mammals and usually close down shortly before or after birth so that the baby's liver takes over the functions of filtration, storage, and production. In some individuals the shunt doesn't close down or develops in an abnormal place, and the animal's liver doesn't get enough blood supply to grow or function properly.

Types of Shunts:

Shunts may be congenital (found at birth) or acquired (developing after birth). Dogs with acquired shunts usually have cirrhosis, or "hardening" of the liver, secondary to severe liver disease. These dogs develop multiple shunting blood vessels to relieve high blood pressure in the liver. There is no effective surgical treatment for these patients, short of a liver transplantation.

Congenital shunts are usually single blood vessels that are present at birth. In large breed dogs, they are found inside of the liver ("intrahepatic") and may be a result of improper or incomplete closure of the fetal shunt.

Surgical treatment of these shunts is possible, but difficult, because of the location of the abnormal blood vessel.

Small and toy breed dogs usually have "extrahepatic" shunts: the blood vessel is located outside of the liver. These shunts are easier to find and treat and therefore the outcome of surgery is better.

Clinical Signs of Portosystemic Shunts

Clinical signs are often seen at a young age and may include poor growth, behavioral changes (circling, disorientation, unresponsiveness, staring into space, head pressing, blind staggers), seizures, and quiet demeanor. Many of the clinical signs may be confused with puppy hypoglycemia (low blood sugar). Other less common signs include diarrhea, vomiting, and excessive drinking or urinating.

In many animals the signs are seen 1-3 hours after eating meat or puppy chow. Proteins in the food are broken down by intestinal bacteria to ammonia and other toxins which are absorbed and, instead of being filtered by the liver, are allowed to reach the brain. The depression and signs are often temporary; once the proteins are emptied from the colon, the signs usually abate. Some animals may not show clinical signs until they are anesthetized to be castrated or spayed. These animals may take days to recover from anesthesia, depending on what drugs were used. Other animals show no signs until they are older, when they develop bladder and kidney problems from excreting toxins and forming urine crystals and stones.

Diagnosis

To diagnose a shunt we may need to rule out toxicity, hydrocephalus ("water on the brain"), and low blood sugar in puppies. We look for abnormalities on bloodwork that indicate poor liver function, such as low protein, albumin, and blood urea nitrogen, which are chemicals produced by the liver. X-rays of the abdomen may show a small liver, indicating that it is has not developed properly. Urine sediment may contain ammonium biurate crystals, which look like starfish or spiky balls.

We also run special diagnostic tests. Blood ammonia concentration can be measured; this test will diagnose liver disease in 90% of affected animals. It is more accurate (95 to 100%) if an ammonia challenge is done, where the puppy is given ammonia by enema or by mouth and the blood is tested to see if the liver clears the ammonia. Blood for ammonia concentration

measurements must be kept chilled and must be analyzed within 30 minutes after it was drawn.

Even more accurate are bile acid concentrations. A blood sample is taken after a 12 hour fast, and then the puppy is fed a normal meal. Two hours later another blood sample is taken. Bile acid concentrations are high in most types of liver disease, including shunts. Bile acid concentrations are altered by hemolysis (breakage of the blood cells) and lipemia (fat in the blood) but are minimally affected by temperature and storage and can be sent through the mail. These tests tell us that liver disease is present but do not verify the presence of a shunt. Bile acids levels that are normal after feeding in a 6 or 8 week old puppy indicate the puppy probably does not have a shunt, as long as there is no hemolysis of the blood sample; therefore this test can be a good screening tool for breeders.

To be 100% sure that a shunt is present, we either need to use ultrasound (which is more useful in large dogs), a contrast study with x-rays ("portogram"), nuclear scans ("scintigraphy"), or surgery to find the shunt. Scintigraphy and ultrasound are not invasive and are therefore the safest tests, with scintigraphy being the most accurate of the two.

Portosystemic shunts must be differentiated from hepatic microvascular dysplasia, a disease that has the same clinical signs, liver biopsy results, and blood changes. This disease is seen in Yorkies, Cairn terriers, and other small breeds and must be ruled out by portogram or scintigraphy.

Medical Treatment

Dogs with shunts that are seizuring or comatose should be given intravenous fluids with dextrose (sugar), intravenous antibiotics, and warm water enemas. Lactulose syrup can be instilled in the rectum after the enema to decrease toxin absorption. Some dogs may even need activated charcoal (given by tube through the mouth into the stomach) to absorb toxins if they are comatose.

Long-term medical treatment for dogs with shunts includes a low protein diet such as Hill's L/D, and drugs such as lactulose and neomycin or metronidazole to prevent production and absorption of ammonia and other intestinal toxins. Lactulose, a sugar solution, is extremely safe, although a high dose will cause diarrhea. Some animals do well on medical treatment alone, but most have shortened life spans because of progressive liver shrinkage and loss of function. Medical treatment is normally used to stabilize a patient until it is healthy enough to tolerate the surgery.

Surgery

Most dogs are taken to surgery to have the shunt closed. Because the liver has not developed properly, many dogs cannot tolerate rapid closure of the shunt. Some veterinarians will partially occlude the vessel, leaving a small amount of shunting present. Many of these dogs still do well after surgery and about half are able to be taken off of medication and special diets within 6 months. In some, the clinical signs may recur, and a second surgery may be required to determine whether further closure can be attempted.

Many veterinarians are now placing ameroid constrictors around the shunts to gradually occlude them. These constrictor rings will slowly close down over 4-5 weeks, allowing the liver to get used to its new blood supply. Survival rates after ameroid constrictor placement are about 95%. Most dogs are completely normal within 3 months. While about 1/3 of the dogs may still have some shunting, less than 15% have any clinical problems.

Postoperative Management

Most dogs will need to be kept on a low protein diet for at least 6 weeks after surgery. Liver function can be assessed at 2-3 months by rechecking albumin and postprandial bile acids, or by scintigraphy. Dogs can be gradually switched to an adult maintenance diet after 8 weeks, but should be kept on a low protein diet and lactulose if clinical signs recur.

Prevention

Portosystemic shunts have been proven to be hereditary in Irish Wolfhounds and Maltese; however, we also see them frequently in other breeds such as Yorkies, Australian shepherds, and Labrador retrievers. Currently I am investigating the pedigrees of Yorkshire Terriers, since the odds of this breed developing shunts are 1,225 times greater than all other breeds combined. At University of Tennessee Veterinary Teaching Hospital, 35% of our shunt patients are Yorkies; at Washington State University, one out of seven Yorkshire Terriers presented to Veterinary Hospital for any reason was found to have a portosystemic shunt. Because of the high prevalence of the disease in Yorkies, we are determined to search for the cause and to look for a "cure".

Latest Update 2002

As many of you know, I have been completing a study funded by the AKC Canine Health Foundation regarding determination of genetic relationships among Yorkshire Terriers with single congenital

portosystemic shunts. Our objectives were to determine whether coefficient of inbreeding was greater in Yorkies with portosystemic shunts (PSS) than Yorkies with normal bile acids (NBA) and to determine whether a common ancestor was present in the pedigrees of dogs with PSS. We excluded pedigrees of full siblings from our analyses so that a wide variety of breedings would be included. Pedigrees were entered into a data collecting system ("The Breeder's Standard"; Man's Best Friend Software, Version 5.0.632, 1999). Wright's coefficient of inbreeding for up to 10 generations was calculated for each dog. Five and eight generation pedigrees were analyzed for common ancestors.

We found that the Wright's coefficient of inbreeding for Yorkies with single congenital extrahepatic portosystemic shunts was twice as high as that of Yorkies with normal bile acids (0.054 +/- 0.094 and 0.028 +/- 0.042, respectively). This difference was not statistically significant, however; we would need to study the a greater number of pedigrees with PSS to see whether inbreeding was a factor in shunt development in Yorkies, as it is in Irish wolfhounds.

We evaluated the pedigrees for common ancestors and, in the PSS group, found one dog that was mentioned repeatedly in most the five and 8 generation pedigrees. However, this dog was also mentioned in most of the pedigrees of the normal Yorkies as well, and therefore cannot be implicated in the disease.

Bottom line: The news is good and bad. For those folks that are concerned about a certain dog or dogs in their lines, there is no proof that a single dog or line is the source of the disease. I'm sure that is a relief for many breeders. However, it would have been much easier to find out that a certain dog or line was responsible, as that would make the problem easier to avoid. Based on our results, the trait is likely to be polygeneic; thus, there is no easy answer as to how to avoid the disease. Veterinarians will likely continue to recommend that neither parent of a shunt dog be bred again or, at the very least, that test breedings of parent to offspring be performed to rule out carriers. Research is now underway at Ohio State University and Michigan to look for a genetic marker. If you know of anyone that has 2 affected dogs in the same line, please contact us or any of the involved Universities. Your information may provide the key to developing a test that will save future generations from this disease.

You are welcome to forward this information to other listservs and internet sites. I'm happy to answer any questions about the study or about shunts.

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Can a Shunt close up on it's own?

There have never been any reports of shunts closing on their own after the puppy is over a week of age. Shunts are permanent problems. Over half of the dogs that have shunts will have progression of their clinical signs and will die or be euthanized in a year unless they have surgery. About a third of the dogs that have shunts can be medically managed for 5 or more years; these are dogs that are older when the diagnosis is made (i.e. several years old) and don't have any neurologic problems (seizures, circling, fly biting, star gazing, blanking out) and have relatively normal blood work (they can make enough protein).