PORTOSYSTEMIC LIVER SHUNT
by: Thomas K. Graves, DVM

LIVER MALFUNCTION IS MORE COMMON THAN YOU THINK
The term portosystemic shunt has a scary sound it, and perhaps rightly so. A portosystemic shunt is a congenital malformation of the veins that bring blood to the liver. Unfortunately, it's not a malfunction you can see with your eyes or hear with a stethoscope or feel with your hands.

What makes it scary is that it's so incredibly vague. It usually affects puppies, but not always. It can be the bottom-line reason for that one that's doing poorly in the litter, but not always. It can be the explanation for unexplained death but not always. It can be the reason the puppy just can't seem to be housebroken, but there can be a million and one other reasons for that. One thing is for sure: It's more common than we think. In a normal dog, blood travels from the intestines through the portal vein to the liver. There it is detoxified and travels to the heart by way of the vena cava. (See above photo.)

In health, the liver's job (among many) is to process the blood. After a meal, blood bathes the intestines, soaking up proteins and their by-products ammonia, for instance) minerals and various other molecules. It collects at a central point in the abdomen and pours into the portal vein. The portal vein then sends this blood which is laden with the by-products of digestion (many are toxic), to the liver.

In the liver, the blood is detoxified before being sent to the vena cava, the large vein that carries blood to the heart. Thanks to this circulatory scheme, the blood arrives in the heart to oxygenated and then to the hungry cells of the brain, muscles and other organs is free of toxic by-products of digestion.

In patients with portosystemic shunt, circulation through the liver is abnormal. There are many different specific malformations that can occur. Most commonly, blood flows from the portal vein directly to the vena cava, bypassing the liver. This is called a portocaval shunt, and can happen with the majority or only a small part of the
When this happens, the blood that is sent to the lungs, brain, cardiac muscles and every other body tissue has not been detoxified. The result is a poisoning of many of the body's cells. Clinically, that can mean just about anything, from poor weight gain to excessive sleepiness and mental dullness to vomiting to blindness to seizures. The longer the shunt goes undetected, the worse the prognosis.

I have seen portosystemic shunts in many breeds of dogs. In my opinion, it occurs commonly in Maltese, Toy Poodles and Shetland Sheepdogs. I've seen it in Labrador Retrievers, Cocker Spaniels, Springer Spaniels, Pugs, Shih Tzu and once in a Corgi. I wouldn't rule it out in any breed, and the fact that it occurs far more commonly in pure-bred dogs than in mixed breeds points to a genetic cause.

THE CLASSIC CASE
The classic picture of a portosystemic shunt patient is that of a puppy. It's smaller than its littermates and has never done well. It sleeps more than usual. It may have a poor appetite and may vomit occasionally and have a little diarrhea from time to time. It may have periods of normalcy, but the pup is at its worst just after eating (that's when toxins in the blood are at their peak). As time goes by the signs of disease become worse. It may progress to periods of poor coordination and blindness. Seizures are common, especially after a meal.

Neurological dysfunction caused by liver insufficiency is referred to as hepatencephalopathy. This can cause any degree of neurological problems including lethargy, dullness, decreased learning ability, weakness, stumbling, blindness, circling, seizure, coma and death.

The last portosystemic shunt patient was not at all like that classical case. It was a two-year old Shih Tzu, a beautiful, typy little bitch with an outgoing personality, a nice topline, four good legs, a very cute face and a ton of coat. The dog was brought to me because of a urinary tract infection. As I discovered the urinary tract infection had been caused by stones in the urinary bladder. The stones were composed of urates, crystalline compounds that build up in the blood when the liver does not function properly. Urate stones form when the levels of urates in the urine are so high as to cause crystals to clump. (See Bladder Stones in Dogs, in September 1992 GAZETTE).

This is the same type of stone commonly found in Dalmatians, but with a different cause. The stones in my patient were due to
inadequate liver function. And the inadequate liver function was due
to a portosystemic shunt. Thankfully, the shunt was surgically
treatable.

If you or your veterinarian have any suspicion of a portosystemic
shunt, the dog should have a full diagnostic workup. The longer it is
left untreated, the worse the prognosis. The liver needs the blood
supply from portal circulation in order to grow and regenerate. In
dogs with severe long standing shunts, the liver is often shrunken and
almost completely non-functional. In such cases, treatment is often
impossible.

The diagnostic starting point (once a thorough history and physical
examination have been evaluated) is a chemistry profile, a complete
blood count and a urine analysis.

Urine is often overlooked as a diagnostic fluid in veterinary medicine.
The reason is the difficulty of obtaining a clean sample. We can't just
ask our patients to go into a bathroom and collect a sample as
aseptically as possible in a little plastic cup. In fact, voided urine
samples in dogs are not very useful. Usually, by the time the urine has
traveled through the vagina or prepuce and through the dog's hair, it
is useless as a diagnostic specimen. And if the urine is collected from
the floor forget it.

I routinely collect urine by cystocentesis; sticking a needle through the
skin on the abdomen and into the bladder. This seems drastic to most
human patients, but is a routine procedure in veterinary medicine and
is safe, almost painless and allows us to get uncontaminated urine
samples from our patients. If I had not collected urine, I would not
have made the correct diagnosis in the Shih Tzu. All of the other tests
on the dog's chemistry profile and blood count were normal.

Changes can be seen in levels of liver enzymes, cholesterol, glucose
and many other factors, but the results of screening blood and urine
tests can only suggest a portosystemic shunt; they can never offer a
definitive diagnosis. To more adequately assess liver function, we do a
serum bile acids test. This test involves taking a blood sample after the
patient has fasted for 12 hours. The dog is then fed and the blood is
retested after two hours.

This tests the liver's ability to clear bile acids (which are secreted by
the liver in response to feeding) from the blood. Any abnormality
suggests liver dysfunction. If post-feeding serum bile acid levels are
normal, this excludes the diagnosis of portosystemic shunt.

**DIAGNOSTIC X-RAYS**

The next step in diagnosis is a trip to the radiology department, where
a variety of tests can be done. I usually start with a set of x-rays of the
abdomen. I'm looking for changes in liver size (the liver can be
smaller than normal in patients with portosystemic shunts). I can also look for bladder stones or kidney stones.

The next step is usually ultrasound of the liver. This test is advantageous because it is non-invasive, perfectly safe and can help rule out other types of liver disease. If it is large enough and if the ultrasonographer is skilled enough, a portosystemic shunt can sometimes be seen with ultrasound.

Another test I use commonly when ultrasound has been inconclusive is rectal portal scintigraphy. It sounds really awful, but it's very safe and usually quite definitive. There is no discomfort or danger for the patient, and the information gathered can be vital. To perform the test, a radioactive substance (with a very low level of radiation) is placed in the dog's rectum via an enema tube. The radiation is taken up by the intestinal circulation and a gamma camera is used to watch the flow of blood by detecting areas of radioactivity in the dog.

In a normal dog, the radiation travels from the intestine to the liver to the heart to the kidneys and beyond. In a dog with a shunt, the radiation travels from the intestine directly to the heart, bypassing the liver. The liver lights up sometime later, and the diagnosis is made. Unfortunately, nuclear medicine facilities are not always available for veterinary patients. However, most university-based veterinary teaching hospitals are equipped to perform rectal scintigraphy.

**Surgery**

The last, and most invasive test is saved for the surgery department. The patient is placed under general anesthesia and a look of gut is isolated through an abdominal incision. A radiographic contrast agent is injected into an intestinal blood vessel and x-rays are taken. The contrast agent allows us to see the path of portal circulation and abnormal vessels can be seen quite easily. Often this test is done at the same time surgical correction of the problem is planned.

There is only one way to cure a portosystemic shunt--surgery. Most veterinarians refer patients with portosystemic shunts to soft tissue surgery specialists. Surgical treatment of the disease involves identifying the abnormal blood vessel (shunt) and closing it. Of course, it's not that simple. It all depends on the degree of normal circulation left and the condition of the liver.

Sometimes, especially in long-standing cases, once the shunt is tied off, there is nowhere else for the blood from the intestines to go. Because there are not enough normal vessels, the blood cannot circulate through the liver and pressure builds up in the intestinal circulation. If this pressure is too high, the patient will die. In such cases, partial closure is attempted.

In some cases, no closure is possible and medical management is the
only option. Medical management can help ease the symptoms, but it is only temporarily effective. It should only be used with patients in which surgery is impossible.

Medical therapy is also valuable before surgery. Depending on the degree of illness, various treatments are used to help decrease toxin production in the gastrointestinal tract and toxin levels in the blood. Low protein diets are an essential part of medical management. Various antibiotics are used to decrease how much ammonia is produced by bacteria in the gut. Lactulose is used to trap ammonia within the intestine and keep it from diffusing into the bloodstream. In severe cases of hepatoencephalopathy, repeated enemas may be needed to reduce blood ammonia levels. Obviously, long-term intensive medical management is difficult, expensive, impractical and, arguably inhumane.

LONG-TERM PROGNOSIS
If a patient survives the first few weeks after surgery, the long-term prognosis is usually excellent. Occasionally seizures that can be extremely severe, develop following shunt closure. The reason for this post-surgical complication is not understood, and the prognosis is very poor. The majority of patients, however, do well following surgery, and they go on to live normal lives.

Like so many diseases of pure-bred dogs, the key to success is early recognition of the problem, aggressive diagnosis and expert care in treatment. The key to prevention is awareness and responsible breeding.

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