

Portosystemic Shunts (PSS)

Portosystemic shunts (PSS) are abnormalities of the liver's blood vessels.

Cause

When abdominal blood vessels develop normally, blood from all the abdominal organs drains through the major liver vessel called the portal vein. The liver filters this blood before it goes to the heart and lungs. The main function of this filtering mechanism is removing the byproducts of protein metabolism, but the liver also has other detoxifying functions. In animals with portosystemic shunts, an anatomic abnormality results in blood being diverted around the liver. This results in blood being circulated without the benefit of the liver's detoxifying function and also deprives the liver of nutrients.

There are a number of different types of portosystemic shunts. Most are congenital and heritable which means that the abnormality was present at birth and has been passed down family lines. Dogs and cats that have portosystemic shunts should not be bred. If your pet was obtained from a breeder, informing the breeder of your pet's condition will help them reduce the likelihood of additional pets being born with this condition.

The most common congenital PSS is a single anomalous blood vessel that is outside of the liver and diverts blood flow around the liver. These are called extrahepatic shunts; they are seen in small breed dogs and are the type most amenable to surgical correction. Less commonly, vessels enter the liver normally, but then divert blood flow in the wrong direction. These are called intrahepatic shunts; they are seen more commonly in large breed dogs and are significantly more challenging to correct surgically.

In some cases, the anatomy of the liver's blood vessels are normal at birth, but a disease of the liver causes scarring and eventually increases pressure in the blood vessels, resulting in the diversion of blood in alternative ways. These are called multiple extra-hepatic shunts. These types of shunts develop in older patients with chronic liver disease and are mostly unrelated to genetics, although some animals have a predisposition to chronic liver inflammation.

Clinical Signs

Depending on the severity, portosystemic shunts can cause a variety of clinical signs. Some patients are apparently normal but have increased liver enzyme tests noted on routine exams. Severely affected patients will have stunted growth, poor appetite, and difficulty maintaining a normal blood sugar from a young age. These patients can become profoundly ill and develop neurologic complications such as confusion and seizures, requiring immediate care. There are also patients with mild signs that go unnoticed until the liver is challenged by anesthesia or disease. Long-standing, small portosystemic shunts can cause bladder stones in some patients that otherwise appear normal.

Diagnosis

Patients suspected of having a portosystemic shunt should have a combination of blood and urine tests, as well as imaging studies. Most commonly, an abdominal ultrasound will be recommended. This study is best performed by an experienced veterinary radiologist using advanced imagery. Vascular anomalies can be difficult to find, so sometimes additional tests, such as computed tomography (CT scan/portogram), often with a contrast agent that makes the abdominal vessels easier to follow, is recommended.

Treatment

Depending on the severity of the patient's condition at the time a portosystemic shunt is diagnosed, hospitalization for intravenous fluids, anti-seizure medications, nutritional and other types of support may be necessary. Once stable, surgical correction is the preferred method for treating most dogs and cats with portosystemic shunts. The goal of surgery is to close the shunting vessel to restore normal blood flow. In order to make patients the best possible surgical candidates, most are put on a special diet and some medications for several weeks prior to surgery. It is important that the pre-surgical protocol be followed to have the best possible outcome.

For patients where surgical correction is not possible, medical therapy is aimed at reducing the workload of the liver through diet and medication to slow the progression of liver failure. Because the liver is responsible for the metabolism of protein, a

low protein diet may be recommended. These diets are generally prescription only as they are not nutritionally complete for all life stages.

An oral medication called lactulose is used to reduce ammonia absorption in the colon. Lactulose is a sugary solution that is not digestible, so it draws water into the colon which can soften the stool. This can be an unpleasant but necessary side effect of this medication. Oral antibiotics are also used to reduce ammonia absorption because they reduce the number of intestinal bacteria that produce ammonia. Anti-seizure medications may be used to combat the neurologic complications of PSS and are used in patients that struggle with seizures.

Surgical correction of the shunt depends on the anatomy. In most extrahepatic shunts, the anomalous vessel is identified and banded by a device that constricts the abnormal vessel over time. These devices were developed to reduce the likelihood that too rapid closure of the vessel would cause complications. Cellophane bands are used most commonly, but a small metal constrictor might be chosen in some patients.

Surgery for single intrahepatic shunts is more challenging. In some cases, it is accomplished through an interventional radiology procedure in which moving radiographs (called fluoroscopy) are used to identify the abnormal vessel. Once identified, the best procedure will be determined. Sometimes a coil apparatus is placed in the abnormal vessel to reduce blood flow. The coil is often placed during the fluoroscopy procedure using interventional radiology techniques rather than surgery. In other cases, the patient needs a surgical procedure to close the vessel. These types of vascular abnormalities can be quite challenging to correct and resolve, but sometimes the procedure reduces flow enough to improve the quality of life and survival.

No surgical treatment exists for multiple extrahepatic shunts. A liver biopsy to diagnose the underlying liver disease will help direct therapy. Biopsy results may help direct appropriate medical treatment and long-term outlook.

Surgical complications are rare but can be significant. In general, dogs handle PSS surgery better than cats do. Because the liver metabolizes anesthetics, every patient at Veterinary Specialty Center has a pre-operative anesthesia evaluation, which significantly reduces but does not eliminate the risk of anesthetic complications. Patients with PSS have diminished protein metabolism, including the proteins involved in blood clotting, so they can have difficulty with healing and require transfusions of red blood cells or plasma proteins from a donor. Because closing off the alternate pathway of blood flow means the liver receives a larger quantity of blood, this can sometimes overwhelm the liver causing a marked increase in pressure called portal hypertension. The incidence of this is lower with newer surgical techniques, but it can still occur.

A serious potential complication of PSS surgery is seizures. These occur most commonly in young patients with a markedly diminished liver function whose developing brains have been negatively impacted by poor liver filtration.

Prognosis

The prognosis for patients with PSS depends on the severity of the anomalous vessels, and whether or not surgical correction is possible. Patients whose shunts are corrected surgically generally enjoy a good long term survival and quality of life with minor restrictions. Some patients with shunts that cannot be corrected surgically do well with medical management for a time; the length of time varies from patient to patient but some patients can be managed for years with good follow-up and monitoring. In patients with multiple extrahepatic shunts due to severe liver disease, the prognosis depends on whether or not the underlying liver disease can be treated effectively.

Long Term Follow Up

The internal medicine doctors at Veterinary Specialty Center manage the patients with portosystemic shunts. Patients with portosystemic shunts that are not amenable to surgical correction are likely to have lifelong challenges. These patients will need to be monitored closely for any progression of liver dysfunction. These follow up visits are likely to include blood tests and, if indicated, imaging studies to assess progress.

Patients that have had a surgical correction of their portosystemic shunt are usually hospitalized under the care of an internist postoperatively. Upon release, each will have specific follow up instructions about medications, diet, and supplements. Virtually all of these patients will remain on a prescription low protein diet for the remainder of their life, but some are able to be weaned off of other medications and supplements within the first year. These changes should only be made if recommended by an internal medicine specialist. Because decisions about changes in medication are based on observations made during the physical exam in addition to other testing, our recommendation is that follow-up for this disease is done at Veterinary Specialty Center. All routine preventive care should continue with your primary care veterinarian. Because patients with portosystemic shunts are unlikely to ever regain full liver function, there will be medication and anesthetic precautions for the remainder of the patient's life.