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State of the Art Care, In the Heart of the State

Porto-systemic Shunt

What are the liver and portal vein and what do they do?

The liver is a remarkable and complex organ located in the very front of the abdomen, behind the diaphragm. The liver is responsible for metabolism and storage of nutrients, processing toxins and drugs, synthesizing proteins and other compounds, and forming and secreting bile to aid in digestion. The portal vein is a blood vessel that brings absorbed digested nutrients from the intestines to the liver for further processing and storage. Blood percolates through the liver before entering into the body as a whole.

- *Bile* is a greenish substance secreted into the small intestine via the gall bladder and bile ducts that contains compounds that aid in digestion of fats; certain drugs or toxins may also be excreted this way
- *Albumin* is the major blood protein made by the liver that helps with a wide array of functions in the body
- *Coagulation factors* are proteins made in the liver that work with platelets and blood vessels to help the blood to clot

What is a porto-systemic shunt (PSS)?

A PSS is an abnormal connection between the portal vein and another normal blood vessel that results in abnormal flow of blood around or through the liver. When a PSS occurs, all the nutrients, toxins, and other compounds that would normally get filtered by the liver enter into the systemic blood supply. These compounds then cause a number of systemic side-effects. PSS are seen mostly in dogs and only rarely in cats.

- *Extra-hepatic* PSS is a shunt that forms outside the liver; this is the most common form, especially in small dogs and cats
- *Intra-hepatic* PSS is a shunt that forms through the liver tissue itself; this is seen more frequently in large breed dogs
- *Microvascular dysplasia (MVD)* is a microscopic form of shunting wherein there are innumerable abnormal connections between tiny blood vessels in the liver; while often seen with PSS, it is a separate disease entity

What causes a PSS?

A PSS can be *congenital* or *acquired*. Congenital PSS may be genetic or due to a non-heritable malformation. These are the most common form of PSS seen in dogs and cats. Acquired PSS develop when blood can no longer flow through the liver as it normally



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would. When this happens, pressure in the portal vein builds up and new blood vessels open, to release this pressure and shunt blood around the liver. This implies that there is some other liver disease or blood vessel problem also present.

- *Breeds* predisposed to PSS include many of the small terriers
- *Congenital* refers to something that an animal is born with
- *Acquired* refers to something that an animal developed after birth

What clinical signs does PSS cause?

While a small percentage of animals with PSS never develop clinical signs, most animals with PSS have a number of clinical signs that can progress without treatment. Most of the clinical signs are associated with the circulation of compounds in the body that cause neurological or behavioral changes. Clinical signs can also develop in association with the low level of albumin made by the liver in animals with PSS. Finally, animals with PSS are predisposed to bladder stones.

Common signs include:

- Lethargy
- Small size and poor growth
- Head-pressing
- Pacing
- Blindness
- Dysuria
- Hematuria

Less common clinical signs include:

- Ascites
- Polyuria and polydipsia
- Seizures

- *Ammonia* and certain protein breakdown products are the main compounds associated with the neurological and behavioral clinical signs; because these levels are highest after meals, clinical signs may worsen at meal times; increased ammonia levels also predispose patients to bladder stones
- *Hypoglycemia* (low blood sugar) is another cause of abnormal behavioral and neurological changes
- *Ascites* is the build-up of fluid in the abdomen; this can be associated with low albumin levels or increased pressure in the portal vein
- *Dysuria* is difficulty urinating, and *hematuria* is blood in the urine; these can result from bladder stones



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- *Polyuria* and *polydipsia* are increased urination and drinking that result from the kidneys inability to effectively concentrate urine in some PSS patients; this is not an indication of kidney disease

What laboratory changes does PSS cause?

PSS can cause many changes on laboratory tests. Ultimately, laboratory tests are required to help confirm the diagnosis of PSS. Laboratory tests can also provide prognostic information.

Common laboratory changes include:

- Low albumin
- Low cholesterol
- Low urea nitrogen (BUN)
- Low blood glucose
- Microcytosis
- Elevated liver enzymes
- Coagulation abnormalities
- Increased bile acids
- Urine ammonium biurate crystals

- *Albumin, cholesterol, BUN* and *blood glucose* are all compounds normally made by the liver; they may or may not be reduced in PSS patients
- *Microcytes* are small red cells typical of PSS
- *Coagulation abnormalities* include predisposition to blood clots or bleeding, depending on what coagulation factors may be deficient
- *Bile acids* are compounds that normally do not enter the systemic blood supply, but rather circulate between the liver, intestines, and portal vein; the systemic levels increase in PSS cases, especially after eating

What testing is recommended for PSS patients?

There are three main goals in evaluating patients with PSS. First, the diagnosis must be confirmed; secondly, therapeutic options must be determined; and finally, prognostic and complicating factors must be evaluated.

Patients evaluated for PSS may need the following tests:

- Chemistry profile
- Complete Blood Count (CBC)
- Urinalysis
- Bile acids test
- Coagulation testing
- Abdominal ultrasound
- Liver biopsy



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Porto-systemic Shunt

- *Bile acids test* is a test that compares bile acid levels before and after eating; levels are usually very elevated, especially after a meal; urine bile acids can also be measured to assess for shunting, although the blood test may be more specific
- *Abdominal ultrasound* is a non-invasive test that uses sound waves to create images of internal organs and structures; this is performed to look for a shunt and to evaluate for underlying disease
- *Liver biopsy* may be done before surgical correction of the shunt with laparoscopy, or at the time of surgery (see below)

What complications can arise in patients with PSS?

Aside from the main clinical signs and laboratory changes, there are relatively few complications of the disease. With both surgical and medical therapy, certain complications can develop. These are discussed below.

What treatment options are available for PSS?

PSS is considered a treatable condition. Because the clinical signs are often very severe, aggressive medical care is required. Medical therapy includes a combination of special diets and medications that reduce circulating toxin levels. Once stabilized, surgery is considered the best option for most patients with PSS. Congenital extra-hepatic PSS are corrected with a surgery in which the surgeon *ligates* the abnormal blood vessel. This returns blood flow to the appropriate course through the liver and can resolve clinical and laboratory changes. At the same time, a biopsy is often collected from the liver to rule-out other underlying liver disease. Alternatively, biopsy can be collected prior to ligation to rule-out the presence of other underlying liver diseases that would preclude ligation.

A small number of patients with PSS still require some form of medical or dietary management after surgery. Additionally, there appear to be a small percentage of these patients with extra-hepatic PSS that have additional liver disease that makes ligation ineffective because acquired shunts develop later.

Patients with congenital intra-hepatic PSS are generally not good candidates for surgery as they tend to have more complications. Newer *coil embolization* procedures may prove useful in the future management of these patients, but are only performed at a few facilities in the country. Acquired shunts should not be fixed with ligation as the underlying disease will cause them to simply re-form.

For patients with intra-hepatic PSS, acquired PSS, or extra-hepatic PSS with other liver disease, long term treatment involves the use of the diets and medications noted above to



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control the clinical signs. Additionally, of course, other specific liver diseases that may be present are treated appropriately.

- *Lactulose* is a medication that controls bacterial ammonia production and reduced ammonia absorption
- *Antibiotics* are used to reduce bacterial load in the intestines and ammonia production
- *Dietary therapy* includes the use of low protein diets to reduce neurological effects
- *Thromboprophylaxis* is the use of medications to reduce the risk of blood clot formation; because patients with PSS may be predisposed to blood clots, aspirin therapy may be used in some cases.

What sort of long-term monitoring is recommended for PSS patients?

Recommendations for follow-up with vary widely depending on whether surgical ligation of the shunt is an option. If surgery is performed, medications will be weaned and CBC, chemistry, urinalysis may be repeated to ensure correction of previously noted abnormalities. Bile acids will likely also be repeated to quantify the success of surgical correction. Many patients do well clinically after surgery, but still may have mildly elevated bile acids. For patients that remain on medical therapy, follow-up will include at least annual CBC/chemistry values along with periodic urinalyses to evaluate for crystal formation. Other recommendations may be made depending on unique aspects of a case.

What is the prognosis with PSS?

The prognosis with PSS varies widely depending on the type of PSS present and treatment elected. Most dogs and cats with extra-hepatic PSS may do very well with ligation surgery and live a full lifespan. Dogs with intra-hepatic PSS and dogs that do not have surgery to correct an extra-hepatic PSS tend to have shortened lifespan, although the quality of life is generally excellent. Animals with acquired PSS have varying prognoses depending on the reason for the PSS. The specifics of your case will be discussed at the time of your appointment.