

CONGENITAL PORTOSYSTEMIC SHUNTS: Wayward vessel(s) challenging to corral.



Overview—“I don’t understand what an portosystemic shunt is; please help me understand the condition and the treatment.”

“Porto-” refers to the portal circulation; the blood circulation specific to a portion of the digestive system.

“-systemic” refers to the whole body circulation

“shunt” refers to a connection, usually an inappropriate connection, between the portal circulation and the whole body circulation. Shunts are typically, but not always, a wayward vessel that used to connect two blood vessel circulations in the fetus that should have gone away at birth, but didn’t.

In this particular case, portosystemic shunts, we are speaking about a vessel or vessels that connect the portal (GI; gastrointestinal) circulation to the venous blood circulation as it heads back to the heart from the body. We refer to it as a “congenital” PSS if it is related to a wayward vessel(s) that started in the young animal, probably the fetus. We refer to them as “acquired” PSS if these vessels developed in the adult related to liver disease and excessive blood pressure in the portal (GI) circulation. *The patients with congenital PSS are the ones we can potentially help with surgery.*

The shunt in these patients causes the portal blood from the digestive track to bypass the cleaning function of the liver and this bypass also prevents the nourishing function of the portal blood from helping grow the liver. The results are the various signs we see in young animals with PSS:

- High levels of blood toxins, such as ammonia, that affect the brain (seizures, weakness, excess salivation, apparent blindness, vomiting, coma)
- Low levels of blood glucose (weakness, coma)
- Inability to thrive (small stature, slow growth)
- Bladder stones from excess urate levels

Dogs and cats can have congenital PSS; the location of the shunt varies most commonly by breed. Small/toy breeds overwhelmingly have shunt vessels “outside the liver” (**extra-hepatic**); larger breeds most commonly have shunt vessels buried “inside the liver” (**intra-hepatic**). These two situations, extrahepatic and intrahepatic PSS, involve different surgical approaches and have different levels of complexity and complications.

“Why is this procedure being recommended for my pet?”

The medical data evaluating what treatment options are best for PSS have mixed information. For the young animal, the statistics suggest surgical closure of the shunt vessel provides the best longterm outcome. For the middle-aged animal with minimal signs, medical management may be statistically the best course. Each individual has its own variables, so careful evaluation is important.

The natural course of untreated PSS is a progressive loss of liver function, an excessive increase in portal blood pressure, and overwhelming insult of natural body toxin to the brain. This process can occur over weeks/months or many years. A small shunt can allow a patient to be free of signs until later in life when bladder stones develop (from an abnormal blood chemistry filtering large amounts of crystal-forming compounds into the urine). Medical treatment can curb the progression of these signs but does not treat the underlying abnormal movement of blood from the portal circulation to the whole body circulation. Most patients with medical management of PSS will not live a normal lifespan, but some can live comfortably, medically managed, for many years.

Surgery closes (or significantly closes) the shunt vessel and eliminates (or reduces) portal blood from bypassing the liver. This restores the liver functions and allows for normal liver nourishment. Life expectancy can be normal for most patients having surgical closure of their shunts.

Diagnostic tests will help define the exact type of PSS a patient has and the status/health of various organ systems it is influencing.

- Blood tests evaluate liver function and overall liver health, blood sugar, toxin levels, etc.
- Urine tests evaluate blood toxin levels, and ironically liver function too.
- Abdominal ultrasound evaluates for bladder stones, and can, with the right hands/eyes/brain, identify the actual shunt vessels inside or outside the liver.
- MRI is another full-abdomen evaluation, like the ultrasound, that can look for the actual shunt vessel and bladder stones.
- Scintigraphy is a useful tool in hard to characterize or postoperative cases that helps define how much blood is bypassing the liver in PSS.

Deciding how which and how many of these diagnostic tests to pursue before choosing a treatment plan is an important step in your decision-making. There are risks to some tests and there are risks in not having more information from those tests. That balancing act is one of the frustrations of medicine!

“What options do I have to treat my pet’s disease?”

Some type and degree of therapy is usually required in patients with congenital PSS to live a comfortable life, though some patients do not develop problems until mid-life. We discuss risks and benefits of these therapies based on patient age & breed, type/location of shunt vessel, liver health, overall health, other medical problems.

Medical management is designed to reduce the overall level of toxins in the blood that are normally cleared by a functioning liver. Dietary protein restriction and several medications can achieve this goal for a period of time such that side-effects (mentally dull, seizures, vomiting) are reduced. Feeding regimens can reduce the impact of low blood sugar. Over time, increasing portal pressure and reduced liver function will result in fluid accumulation in the abdomen. Medications and diet will lose their effectiveness in reducing toxins that effect the brain (dullness, seizures). Quality of life will deteriorate. This process can be rapid or develop over several years, and it is very difficult to predict which course each patient will take.

Surgery is undertaken to identify the shunting vessel and reduce or tie-off its blood flow; this results in the return of blood flow directly to the liver. There are two major difficulties with surgical treatment: 1) identifying the abnormal vessel and 2) reducing blood flow through the shunt without suddenly creating excessively high and life-threatening portal pressure. Diagnostic tests and surgical experience help with the first issue of finding the shunt vessel, though in some patients (especially with shunt vessels inside the liver) this is extremely difficult. The second issue is managed by reducing the shunt vessel over time with slowly compressing devices applied to the vessel or reducing the vessel only partially. Surgical experience directs decision-making regarding shunt vessel treatment but risks always remain.

“What postoperative complications do I need to know and understand when considering this surgery?”

Immediately during or after surgery, the complications that may develop and cannot be predicted are:

- Portal hypertension (sudden, life-threatening, requiring emergency surgery to undo the shunt treatment)
- Poor anesthetic recovery (potentially life-threatening, requiring critical care support)
- Seizures (uncommon, develop 1-5d postoperatively, require critical care support and medical coma)

- Blindness (uncommon, rarely reversible)
- Death

Longer term, complications include problems related to either persistent shunting through the original vessel, or the development of new shunts in response to closure of the original. These are:

- Bladder stone formation
- Return or persistence of dull mentation, seizures, low blood sugar, vomiting
- Development of abdominal fluid accumulation (ascites)

“How is my pet’s life and lifestyle likely to change after this procedure?”

For patients dependent upon medications for adequate quality of life, surgical closure of a shunt vessel can return them to a lifestyle without medications, or reduced medications. Most patients will improve their appetite, growth and development, and mental alertness and personality.

“Are there things I can do to prepare myself, my home and/or my pet for this procedure?”

Preventative seizure medications can be started 1-2wks prior to surgery and continued several weeks after surgery. Please secure a prescription from your pet’s primary care veterinarian and administer according to directions.

Medications and special diet to control the signs of excess toxins should be administered until surgical treatment is completed and the liver has had time to recovery (several weeks.) Please secure prescriptions and recommendations from your pet’s primary care veterinarian and administer according to directions.

Access to a full service, critical care veterinary hospital is essential for emergency complications. Please prepare accordingly.

In-patient surgery with onsite critical care is an option that should be considered when making decisions about treatment for your pet.

Outpatient surgery and anesthesia can be uncomfortable, painful, disorienting, and frustrating experiences for animals; watching your pet work through the early postoperative period and recover from anesthesia and pain medications can be worrisome, scary and frustrating for pet owners. The majority of the time this period of difficulty is brief, and *your pet is actually more comfortable and secure at home with you*. Sometimes it doesn’t feel like that at two in the morning when your pet is anxious and not consolable, and you are unsure of what to do. You always have the option of transporting your pet to a 24-hour veterinary facility postoperatively. If you do not want to have your pet home in the first few days postoperatively, please advise your primary care veterinary staff. They will provide contact information for a local 24-hour veterinary facility and help get an estimate for the ongoing care.

It is important that you have proper expectations about this procedure; your experience and you pet’s outcome will benefit greatly. Please discuss this information with your veterinarian when working through the decision-making process regarding **Portosystemic Shunt**.

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