Portosystemic Shunt (PSS)

Authored by: Becky Lundgren, DVM

A portosystemic shunt (PSS, portasystemic shunt, portocaval shunt, portacaval shunt, liver shunt, hepatic shunt, or porto-systemic vascular anomaly) happens when a pet’s venous blood from the stomach, intestines, pancreas, and spleen bypasses the liver. The pet can be born with the shunt (congenital) or can get it later (acquired). About 80% of the cases are congenital, although sometimes a pet can have both types. About 25-33% of the congenital shunts are within the liver. Approximately 0.18% of all dogs have congenital shunting.

During pregnancy, the portal blood vessel in the fetus bypasses the liver (the mother’s liver filters out toxins for the fetus). Normally this shunt closes within three days after birth. In affected animals, the shunt doesn’t close and the blood continues to bypass the liver. Because the liver filters toxins, if it is bypassed the toxins build up in the body. This results in the puppy or kitten having slow or nonexistent growth (failure to thrive). If left untreated, puppies and kittens are not likely to survive.

Congenital shunts seem to happen more in purebreds than in mixed breeds. Breeds with increased risk of PSS include Yorkshire terrier, Maltese terrier, Silky terrier, miniature schnauzer, miniature and toy poodles, Lhasa apso, Bichon Frise, shih tzu, Havanese, Dandie Dinmont terrier, Pekingese, German shepherd dog, golden retriever, Doberman pinscher, Labrador retriever, Irish setter, Samoyed, Old English sheepdog, Irish wolfhound, Australian shepherd, Australian cattle dog, Himalayan, and Persian.

No sex predisposition has been documented.

An acquired liver shunt is usually caused by liver problems (hepatic cirrhosis, portal hypertension, hepatic arterio-venous malformations, etc.) that resulted in the body routing blood through whatever blood vessels are available, even if it means bypassing the liver. (It would be like taking side streets to your final destination, instead of using the interstate highway.) As happens with congenital shunts, the liver can't filter what doesn't pass through it, so toxins build up in the body.

Signs

Signs include stunted growth, not gaining weight, losing weight, vomiting, diarrhea, lethargy, unresponsiveness, temporary blindness, seizures, spaciness (staring into space), disorientation, circling, poor skin and coat, excessive drinking, excessive urination, etc. Sometimes the pet will just act odd after eating, or pace around or press his head against the wall. The signs you will see depend on the location of the shunt and how many toxins have built up in the body. Some pets will only have one sign, while others could have several. Many of the clinical signs associated with PSSs are related to hepatic encephalopathy.

Diagnosis

Clinical signs, blood tests, urinalysis, and imaging tests (e.g., radiographs, ultrasound images, portograms [an image of the blood vessels to the liver], or nuclear scintigraphy [a nuclear scan that measures blood flow]) can be used for diagnosis. Blood bile acids are elevated after a meal, so the before-meal and after-meal bile acid levels are compared. Despite the variety of tests available, a confirmed diagnosis may not be available until surgery is done.

Treatment
Some pets with liver shunts can do well with medical management. However, some of those pets will eventually be euthanized when dealing with the neurologic problems (hepatic encephalopathy) becomes too much for an owner to cope with. In some pets, a change of diet can be enough to control the signs. A typical diet would involve low-protein, low-magnesium, high zinc, and high Vitamin E, in addition to lactulose. Medical management is more a matter of avoiding certain things (diuretics, NSAIDs, barbiturates, infections, etc.).

Surgical repair of the shunt is common, particularly for congenital shunts, but it seems to be more successful in dogs than in cats. Before surgery, the veterinarian will try to stabilize your pet as much as possible with a low-protein diet and antibiotics. Post-operative antibiotics will also be given. After surgery, once the pet has normal bile acid levels, he can usually return to a normal diet.

It takes about two weeks after surgery before the pet feels better. During that time, many pets will be off their feed. Owners may have to coax their pets to eat, feed canned or strained meat diets, etc. Ask your veterinarian about stimulating your pet's appetite.

**Monitoring**

Medical management of hepatic encephalopathy (e.g. antibiotics, lactulose, protein-restricted diet) will be continued for several weeks following surgery or until hepatic function improves. Laboratory tests are usually re-evaluated one month after surgery to check for evidence of improved liver function. Bile acids are usually re-evaluated three months after surgery. If they are still abnormal, then medical therapy is continued. If bile acids are still elevated six months after surgery, your veterinarian will re-evaluate your pet for persistent shunting. Many dogs with repaired intrahepatic shunts can have persistently elevated serum bile acids. Even with abnormal bile acid levels, the absence of clinical signs and the normalization of other blood chemistry parameters are considered markers of a good outcome.

**Prognosis**

How well the pet does with medical management or surgery depends on many factors, such as where the shunt is located, if the pet has both acquired and congenital shunts, etc.

Approximately 33% of patients with PSSs can be managed with medical therapy alone. The older the animal is at the time of onset of clinical signs, the longer their survival with medical management alone.

The overall success rate from surgery is about 85%. Complete surgical ligation of the shunt has a good prognosis. A partial ligation will provide some level of improvement, but will have a less positive long-term prognosis. Some animals will not respond to any treatment, and will have to be euthanized.

Treated cats don't do as well as treated dogs. Sometimes cats will still have neurologic signs after surgery and may need continuing medical treatment. Some cats may respond to a second surgery one month after the first one. Only a third of the cats who have the shunt ligation surgery do well long term.

With dogs, the prognosis really depends on where the shunt is located.

Unfortunately, over half of the dogs who don't have surgery and are treated medically are eventually euthanized, typically within 10 months of diagnosis. This is because of neurologic difficulties or continued liver damage. Dogs that do best with long-term medical management are usually older at the time of diagnosis and aren't as severely affected. Surgery is usually, but not always, the best option for a normal life span.

Affected pets should never be bred because it is too much strain on the body and because this condition runs in families.