

## **CONGENITAL PORTOSYSTEMIC SHUNTS: THEY ARE REALLY COMMON AND THE TEXT BOOKS ARE WRONG**

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Congenital portosystemic shunts (pss) are much more common and certainly much more confusing than we ever imagined. At Texas A&M, we infrequently see the “classic” congenital PSS with the relatively straight forward presentation (i.e., young Yorkie with post prandial hepatic encephalopathy), probably because those cases are efficiently filtered out and never referred to us. Some breeds are more commonly affected (i.e., Yorkshire terriers, Pugs, Maltese, Schnauzers, Poodles, Shih Tzus, Havanese, Irish Wolfhound, Golden Retrievers, and Labrador Retrievers), but any dog may have a congenital PSS. We infrequently see classic post-prandial hepatic encephalopathy; rather, we more commonly see a young dog (e.g., one of the above breeds that is less than a year old) that is a “poor doer” who is not as big or as strong as the litter mates with very intermittent vomiting (i.e., “he or she has always had a sensitive stomach”) and subtle signs of encephalopathy. Therefore, it is important to eliminate intestinal parasites and hypoglycemia in animals with suspected congenital PSS since the signs may be very similar. Polyuria-polydipsia can be a major clinical sign. In fact, in our practice, most young animals referred for possible central diabetes insipidus turn out to have hepatic disease, especially congenital PSS.

Classic hepatic encephalopathy consists of post-prandial seizures, coma, somnolence, blindness, head pressing and/or aggression. However, we are seeing more and more animals in which hepatic encephalopathy is manifested simply by their laying around a lot, acting tired or lethargic, or just not being interested in anything. In many cases, there is no obvious relationship between eating the signs. In some cases, about all you can say is that the patient has always been a “calm” dog and never really caused a lot of trouble by getting into things. In older dogs, the only comment by the owner may be that the dog is “getting older and slowing down a bit”. To make matters more confusing, we are finding dogs that have hepatic encephalopathy that do not respond to medical management with lactulose or metronidazole. Some of these patients only quit having signs of hepatic encephalopathy when the shunt is surgically corrected. Therefore, you cannot allow lack of response to medical therapy help you decide whether or not a dog has hepatic encephalopathy due to a congenital PSS. Cats with hepatic encephalopathy due to congenital portosystemic shunting often have drooling as a major presenting complaint.

We sometimes see hematuria due to ammonium urate urolithiasis, but this usually often happens in older dogs (especially Schnauzers) that have had chronic hyperammonemia. Many times, this is the only clinical sign in the affected patient. Contrary to what is often described in textbooks, you can sometimes see major increases in ALT and SAP. We occasionally see patients with major increases in ALT (i.e., > 1,000 U/L) that appear to have acquired hepatic disease, probably toxic in nature. The ALT waxes and wanes with clinical signs. Our guess is that these dogs only have signs when they develop liver disease secondary to exposure to “toxins” that the atrophied liver cannot process because it is insufficient.

To further complicate the situation, we are seeing more and more dogs with congenital PSS that are being diagnosed for the first time when they are 7 or even > 10 years old. This appears to be especially common in Schnauzers, although other breeds may also be affected. Many times

these patients have relatively minor signs that have been considered as normal for the particular patient (i.e., has always been a quiet dog, has always been a smallish dog, etc).

Ascites is exceedingly rare in animals with congenital portosystemic shunts. This is in distinction to dog with congenital hepatic AV fistula, which is another congenital vascular abnormality but which is entirely different from the standpoint of signs, diagnosis, and treatment. Ascites is relatively common in dogs with acquired portosystemic shunting. Therefore, if ascites is seen, one should first look for other hepatic diseases. In like manner, icterus is very seldom caused by congenital portosystemic shunts, and finding hyperbilirubinemia is an indication to first look for other diseases. In summary, congenital PSS present in a variety of ways, many of which are not the “classic” presentation that is described in textbooks.

The major criteria for presumptive diagnosis of congenital portosystemic shunts has classically consisted of an appropriate history and physical examination as well as obvious microhepatia and very increased serum bile acid concentrations. It was generally anticipated that dogs with congenital PSS would have serum bile acid concentrations  $> 90$  mmol/L. Hypoalbuminemia, hypocholesterolemia, and/or decreased BUN are common findings on clinical pathology, but they are not invariable; some patients with congenital PSS do not have any abnormalities on the serum biochemistry panel. Ammonium biurate crystals in the urine are useful if they are present; but, most of the cases of dogs with congenital portosystemic shunts that we see do not have ammonium biurate crystals in the urine.

It now appears that serum bile acid concentrations are not as easy to interpret or as definitive as many people think. First, you must always measure both resting and post-prandial concentrations because about 20% of dogs have a resting serum bile acid concentration that is higher than the post-prandial serum bile acid concentrations. Second, there can be marked variation in serum bile acid concentrations from day to day. It is easy to see a two-fold difference in values taken a few days apart, and we have seen a three-fold increase in samples that were taken 72 hours apart. Third, some dogs with congenital portosystemic shunts have surprisingly low serum bile acid concentrations. We have found dogs with congenital PSS that have what we would consider relatively modest increases in serum bile acids (e.g., 55-65 mmol/L, which is a value found in many animals with clinically insignificant hepatic disease), and rare cases have completely normal serum bile acid concentrations. In distinction, some dogs without any demonstrable hepatic pathology other than vacuolar hepatopathy have values in excess of 200 mmol/L. This major overlap in the values of serum bile acid concentrations in dogs with and without clinically significant hepatic disease leads to diagnostic confusion in some cases.

Hyperammonemia is very specific for hepatic insufficiency, especially congenital PSS. However, it is easy to have laboratory artifacts that falsely increase these values. This test can only be run in house, and the instructions must be followed to the letter to avoid artifactual results. Measuring only fasting blood ammonia concentrations is approximately 80% sensitive for congenital PSS (and lower for diseases causing acquired hepatic insufficiency). The ammonia tolerance test is an excellent test with very high sensitivity and specificity, but it is a royal pain to do (e.g., would you like to drink ammonia chloride or have it infused into your

rectum?) and consequently is seldom performed. Measuring blood ammonia concentrations 4-8 hours post-prandially seems to enhance the sensitivity for congenital PSS up to about 90%.

Imaging can be helpful, but one must recognize the limitations of these techniques. We expect to see microhepatia in dogs with PSS, although sometimes the change is very modest. Sometimes there is a marked difference in the apparent size of the liver on the left lateral versus the right lateral projection. Radiographs are a much more sensitive way to find microhepatia than ultrasound. If there is any doubt about the size of the liver, one can administer a few mls of barium sulfate to help outline the stomach, allowing one to easily ascertain the cranial border of the stomach. The area between the cranial border of the stomach and the diaphragm is usually the liver. However, occasional animals will appear to have a small liver when in fact they have a normal sized liver. Fortunately, this situation appears to be unusual and should be picked up if lateral and DV views are obtained.

Ultrasound is commonly employed when looking for congenital portosystemic shunts. A good ultrasonographer can find a congenital PSS about 50-75% of the time, if they are accomplished and can take their time and look. Truly exceptional ultrasonographers seem to find congenital PSS about 90% of the time. Therefore, you must remember that failing to find a congenital PSS on ultrasound does not eliminate it. Furthermore, one cannot look at the liver to see if there are apparently normal portal areas as a means of deciding if a congenital shunt is more or less likely. We have seen animals with congenital PSS that appeared to have normal portal vasculature on ultrasound, to the point that the conclusion was that a congenital shunt was very unlikely. Ultrasonography is a very good way to check for an intrahepatic shunt, which is much harder to correct than an extrahepatic shunt.

Other imaging techniques may include operative or percutaneous portograms, nuclear scintigraphy, and MR or helical CT. These latter techniques should primarily be done for one of two reasons. First, the case is "atypical" and it is important to absolutely confirm the presence of a congenital PSS before going to surgery. The second reason is that the surgeon is unable to find the shunt during an exploratory laparotomy. In general, it is not always necessary to definitively "see" the shunt via some imaging modality before going to surgery. If the case is classic in that it is a young animal with appropriate signs and an obviously small liver and obviously high serum bile acids or ammonia, then one is justified in going to surgery even if the shunt has not been visualized. If the shunt cannot be found during surgery, then an intraoperative portogram can be performed. However, if any of those three criteria are not met (i.e., "classic" history, obviously small liver, obviously increased serum bile acids or ammonia), then confirmation by portography, scintigraphy, CT or MR is appropriate.

Retrograde portography is often preferred when an intrahepatic shunt is believed likely because we prefer to fix these with catheters (i.e., putting in a stent and then coils). Nuclear scintigraphy is also very nice, but requires special facilities. One advantage of portography is that one may place the catheter in the shunt and leave it there in order to help the surgeon find the shunt if they are having a very difficult time finding it.

Lastly, it is important to do a full work up (i.e., CBC, serum chemistry panel, abdominal radiographs, abdominal ultrasound, serum bile acids or blood ammonia) on all dogs with

suspected congenital PSS. These dogs may have other, concurrent diseases. In fact, dogs with previously well compensated congenital portosystemic shunts may not become symptomatic until another disease process causes the patient to start showing signs due to the shunt. Furthermore, a reasonable number of affected dogs have cystic calculi that can be removed during the surgery to correct the congenital shunt.

Surgical correction is usually preferred for younger animals and for those that have signs of encephalopathy that are not controlled with medical therapy. But, surgery is not without risks. The Ameroid constrictor makes the surgery much easier and quicker than before. However, about 15-20% of dogs that have surgery to correct a PSS will have some post-operative complications (usually something minor like ascites). This is usually not a major problem, but the owner needs to be warned ahead of time. Some dogs develop enough portal hypertension to cause acquired PSS, and a few (i.e., 5-7%) have major, life-threatening problems (e.g., post-ligation seizures, portal hypertension) and die. Not every dog with a congenital PSS is benefitted by Ameroid constrictors.

A major concern centers around dogs (especially those 5 years old and older) with congenital PSS that are clinically normal and that have minimal changes on serum biochemistry panel and a liver that is not too small on radiographs. We are finding these dogs because awareness of congenital PSS has substantially grown, and more and more people are looking for them and diagnosing them in animals with minimal or even no clinical signs. If the liver is not too small on radiographs, the serum albumin is  $> 2.0$  gm/dl, and there are minimal to no clinical signs, then we might decide to watch them to see if they will ever need surgery. Dogs with congenital PSS causing hepatic encephalopathy typically benefit from corrective surgery. There is concern that dogs  $> 5$  years of age are more likely to have severe complications from corrective surgery. While this might be the case, many dogs have benefitted from surgery despite being  $> 5$  years of age. This entire area is currently very controversial. We see some dogs with congenital PSS that seemingly live a normal life and never need corrective surgery. Therefore, if you are considering surgery in an older dog (e.g.,  $> 6$  years old) without any major clinical signs, you should probably have a long talk with the owners about how the dog could be worse after the surgery than it was before.

If post-ligations seizures occur, you must first be sure that the dog is not hypoglycemic. The cause of this problem is uncertain, but some suggest it might be due to cerebral edema. We have not treated for cerebral edema in these patients; rather, we typically anesthetize them with a constant rate infusion of propofol until the seizures have stopped. Do not use diazepam or phenobarbital. Some people recommend treating dogs with potassium bromide or Keppra and cats with phenobarbital before surgery for congenital portosystemic shunts, in an effort to avoid this problem. This approach is contentious, and time will tell if it is correct or not. In general, cats with congenital portosystemic shunts do seem to have more post-operative problems than dogs.

Dogs with intrahepatic shunts have a worse prognosis because the surgery is technically much more difficult to perform. If you can refer the dog to a center which can place coils in the shunt via intravenous catheters used with fluoroscopy, that might be a much safer way to try to correct the problem.

The medical treatment for hepatic encephalopathy is relatively straightforward; lactulose, metronidazole, and a low protein diet. However, the concept of low protein must be revisited. Giving too little protein is extremely detrimental to the liver. The goal is to give as much protein as the liver can tolerate. In particular, it is best to give milk and vegetable proteins instead of meat proteins.