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CONGENITAL PORTOSYSTEMIC SHUNTS: DIAGNOSIS AND TREATMENT IS MORE COMPLEX THAN WE THOUGHT

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Abstract

Congenital portosystemic shunts are more common than many people realize. The clinical presentation varies much more than many people realize. In particular, treatment of older patients is not always obviously surgical.

Introduction

Congenital portosystemic shunts (PSS) are much more common and certainly much more confusing than we ever imagined. 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. Polyuria-polydipsia can be a major clinical sign.

Hepatic encephalopathy

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 he 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 they dog is “getting older and slowing down a bit”.

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.

**Older patients**

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). In summary, congenital PSS present in a variety of ways, many of which are not the “classic” presentation that is described in textbooks.

**Serum bile acids**

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. It now appears that serum bile acid concentrations are not as easy to interpret or as definitive as many people think. There can be marked variation in serum bile acid concentrations from day to day. It is easy to see a two to three-fold difference in values taken a few days apart. 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. Measuring only fasting blood ammonia concentrations is approximately 80% sensitive for congenital PSS (and lower for diseases causing acquired hepatic insufficiency).

**Imaging**

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. 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. 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.
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 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.

**Treatment of older dogs**

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 may benefit from corrective surgery, but some do not. 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.

**Selected readings:**


