Advances in congenital portosystemic shunts

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ABSTRACT

A congenital portosystemic shunt (CPSS) is an abnormal vessel between the hepatic portal circulation and the systemic circulation, bypassing the liver. This is a rare condition in dogs and cats, but frequently diagnosed nowadays.

Medical history, signalment, clinical signs, and biological and diagnostic imaging results will diagnose a CPSS and rule out other hepatic or neurological diseases.

Medical management is a turning point to treat and stabilise the animal. Surgical attenuation of CPSS is recommended to improve long-term outcome in dogs and cats, but at least two weeks of medical management is required before surgical treatment is attempted. Surgical options include acute ligation with suture, gradual occlusion with ameroid constrictors, cellophane banding or hydraulic occluders, or embolisation with coils. Excellent outcomes have been described in 80% to 85% of dogs undergoing gradual shunt occlusion and in more than 87% of dogs undergoing coil embolisation.

This article focuses on different biological and diagnostic imaging methods to confirm a CPSS, and outcome in dogs and cats undergoing surgical shunt attenuation or medical management alone.

A congenital portosystemic shunt (CPSS) is an abnormal intrahepatic or extrahepatic vessel that allows blood from the hepatic portal circulation to drain directly into the systemic circulation, bypassing the liver1-4.
Development of congenital defects may be influenced by environmental or genetic factors, or in combination. A genetic influence is suspected when the prevalence of disease is higher in a breed.

Congenital portosystemic shunts have been reported in 0.18% of all dogs and 0.05% of mixed-breed dogs. Breed association has been proven in the Havanese (3.2%), Yorkshire terrier (2.9%), Maltese (1.6%), Dandie Dinmont terrier (1.6%), pug (1.3%), Skye terrier (1.2%), miniature schnauzer (1%), longhaired Chihuahua (0.9%), Scottish deerhound (0.9%) and schnauzer (0.8%).

Intrahepatic shunts (Figure 1) are over-represented in larger breed dogs, with a prevalence in the Irish wolfhound, Labrador retriever, golden retriever, Australian cattle dog and Australian shepherd dog. In cats, extrahepatic shunts are more commonly identified and reported in domestic shorthair, Persian, Siamese, Himalayan and Burmese breeds.

Clinical signs

Dogs with a CPSS often grow poorly or have clinical signs related to the nervous, gastrointestinal or urinary systems. However, cats present with non-specific or vague clinical signs.

Most cats (93% to 100%) present with some degree of neurological abnormality. Hypersalivation is a hallmark of CPSS in cats, reported in 67% to 84% of cases. This neurological sign is caused by hepatic encephalopathy and reflects a porencephalic localisation. In addition, 13% to 64% of cats with CPSS are reported to have copper-coloured irises.

Differential diagnosis
Single CPSS must be differentiated from other neurological conditions (hydrocephalus and epilepsy) and from other primary hepatic diseases, including congenital portal vein hypoplasia with secondary microvascular dysplasia (PVH-MVD) and multiple acquired shunts secondary to portal hypertension.

Differentiation is usually based on history, clinical signs, results of blood work and advanced imaging. Results of scintigraphy, portography and CT angiography are usually normal in dogs with PVH-MVD. Unlike animals with CPSS, ascites is common in dogs with portal hypertension secondary to severe hepatocellular disease or hepatic arteriovenous malformations.

# Diagnosis

## Biochemistry and haematology

In dogs, common blood work abnormalities include microcytosis and decreases in blood urea nitrogen, total protein, albumin, glucose and cholesterol. Alkaline phosphatase and alanine aminotransferase levels are normal or may be moderately increased, whereas total bilirubin is in the normal level.

A rise in alanine aminotransferase has been suggested due to reduced hepatic perfusion, leading to hypoxic cellular damage. However, a rise in alkaline phosphatase is most likely from bone growth.

Cats with CPSS may have normal albumin, glucose, total protein and cholesterol concentrations, but usually have a moderate increase in liver enzymes. The most common abnormality is low urea, seen in 61% to 100% of cats with a CPSS.

## Urinalysis

Urinalysis may reveal low urine-specific gravity, particularly in dogs and cats with polyuria or polydipsia. Other urine abnormalities include ammonium biurate crystalluria and, occasionally, abnormal urine sediment suggestive of cystitis secondary to crystalluria or (urate) urolithiasis. In cats with CPSS, ammonium biurate crystalluria and calculi are reported in 20% to 43% and 12% of cats, respectively.

## Hepatic function tests

Two blood tests are commonly used – blood ammonia and the bile acid stimulation test.

Fasting ammonia is increased in most cats with CPSS; however, normal ammonia level should not
rule out diagnosis. The sensitivity and specificity of ammonia for CPSS diagnosis has been reported as between 91% and 84% in dogs, and 83% and 86% in cats, respectively.\(^3\)\(^,\)\(^7\)

A bile acid stimulation test is the most accurate test to evaluate liver function for the diagnosis of hepatobiliary disease. Bile acid concentrations are usually greater than 75?mol/L in dogs with CPSS, but also with any significant liver disease. Postprandial bile acid concentrations are, occasionally, less than the prefasting sample (in approximately 20% of dogs) because of spontaneous interdigestive gallbladder contraction.

The sensitivity of preprandial and postprandial bile acids has been reported as 58% to 100% and 100%, respectively, with a specificity of 84% for fasting bile acids.\(^3\)\(^,\)\(^6\)

**Diagnostic imaging**

**Radiography**

Abdominal radiographs may be performed as part of a diagnostic work-up, but add little to the diagnosis of a CPSS. Common findings on radiographs include a small liver and enlarged kidneys. Urate calculi are not visible unless combined with other compounds, such as struvite or calcium.\(^2\)\(^,\)\(^3\)\(^,\)\(^6\)

**Ultrasonography**

CPSS can be identified on ultrasonography by experienced operators, using colour-flow Doppler. Dogs and cats with extrahepatic shunts have reduced portal vein-to-aorta ratios, small liver, enlarged kidneys and uroliths. This procedure is extremely useful, relatively inexpensive and non-invasive, but highly operator-dependent.

Ultrasonography has been reported to have a sensitivity and specificity of 100% in one study, but other studies have allowed an identification in only 47% to 50% of CPSS in cats.\(^2\)\(^,\)\(^3\)\(^,\)\(^6\)

**Portovenography**

Direct injection of contrast into the mesenteric, splenic or jejunal veins (portovenography) usually provides excellent information regarding the presence, number and location of shunts. However, some CPSS are not visible on portograms performed in dorsal or right lateral recumbency.\(^3\)\(^,\)\(^6\)

**Intraoperative portovenography**

Intraoperative portovenography using fluoroscopy allows the imaging to be performed at the same time as surgical attenuation of the shunt. Thus, this procedure can also be used to assess the intrahepatic vasculature and be used as a prognostic indicator. Dogs and cats with well-developed intrahepatic vasculature are likely to show a better response to treatment, with fewer complications.\(^3\)\(^,\)\(^6\)\(^,\)\(^8\)
Nuclear scintigraphy

Nuclear scintigraphy with technetium-99m provides a diagnosis of shunt or no shunt, but limited anatomical information regarding the shunt, making it less useful for preoperative surgical planning. Its use is limited by radiation safety guidelines and availability. However, it may help diagnose more complicated or occult vascular anomalies and in the follow-up of surgically treated animals to assess for continued shunting.3,6,9

CT angiography

CT angiography is considered the standard for definitive diagnosis of CPSS and detailed information regarding shunt morphology and intrahepatic vasculature development before surgery. One study found CT angiography was 5.5 times more likely to correctly ascertain the presence or absence of CPSS than abdominal ultrasonography.3,6,10

Magnetic resonance angiography

Magnetic resonance angiography can also detect shunts, but it is more expensive, it takes longer and image quality is not as good as those obtained by CT angiography.3,6

Medical management

All animals with CPSS should receive medical management to improve their physical condition, blood results (albumin and total protein levels) and treat or prevent hepatic encephalopathy (HE).

A minimum of two weeks’ medical management before attempting surgical treatment is recommended.3,6

Dietary protein is restricted moderately to reduce substrates for ammonia formation by colonic bacteria. Crude protein requirements in dogs with liver disease are approximately 2.11g per kilogram of bodyweight per day. On a dry matter basis, commercial liver diets range from 14% to 18% protein for dogs and 31% to 32% for cats. Soybean meal and dairy proteins are highly digestible, so are recommended.6

Lactulose, a synthetic disaccharide, is hydrolysed to organic acids by enteric bacteria in the colon. This acidification of colonic contents results in the production of more ammonium (NH+4) than ammonia (NH3). Ammonia is freely diffusible across the mucosa, whereas ammonium is not. Hence, acidification reduces the amount of ammonia absorbed.

Dosage should be regulated so faeces are soft, but formed. A common dose in dogs is 0.5 ml/kg to 1ml/kg orally every 8 hours to 12 hours. In cats and toy breed dogs, a common dose is 0.5ml to 2ml orally every 8 hours to 12 hours. Lactulose can be given by enema in obtunded or seizing
Antibiotics reduce the enteric flora that produce many of the toxins implicated in HE during digestion of a meal. Ampicillin, amoxicillin, amoxicillin/clavulanic acid, neomycin and metronidazole have all been used to treat HE in dogs and cats\(^3\).

**Figure 2.** An ameroid constrictor can be used to occlude the shunt.

Antiseizure medication should be started in dogs and cats with neurological signs that fail to respond to medical therapy for HE, or those with uncontrolled seizures or status epilepticus. In a retrospective study, postoperative seizures were reported in 4 of 84 dogs that did not receive preoperative levetiracetam before CPSS attenuation, compared with 0 of 42 that received preoperative levetiracetam\(^1\). Moreover, levetiracetam has a more rapid onset of action and no liver toxicity in comparison to phenobarbital.

In dogs and cats, a common dose of levetiracetam is 20mg/kg orally every eight hours\(^3,6\).

**Surgical management**

Once patients have been managed medically for several weeks, attenuation is recommended to improve long-term outcome.

Surgical options include acute ligation with suture, gradual occlusion with ameroid constrictors (Figure 2), cellophane banding or hydraulic occluders, or embolisation with coils (most commonly for intrahepatic CPSS)\(^3,6,12,13\).
Prognosis with medical or surgical management

With medical management, weight and quality of life stabilises or improves in most animals. However, long-term mortality rates of medically treated animals are higher than those of animals undergoing shunt attenuation\textsuperscript{1,6,14}.

Survival time of cats with CPSS, treated with medical management alone, is less than two years\textsuperscript{3}.

Dogs and cats with multiple acquired CPSS are managed medically\textsuperscript{1,3,6}. Complications and mortality rates are highest in animals that undergo shunt ligation; therefore, gradual occlusion or coil embolisation is preferred. Excellent outcomes have been described in 80% to 85% of dogs undergoing gradual shunt occlusion and in more than 87% of dogs undergoing coil embolisation, as long as the dogs are maintained on lifelong antacid (for example, omeprazole) therapy to prevent gastrointestinal ulceration\textsuperscript{1,6,14}.

Good or excellent long-term outcome is reported in about 75% of cats that survive surgery. The most common postoperative complication is neurological dysfunction, including generalised seizures and central blindness in up to 28% and 44% of cats, respectively. Blindness usually resolves in two months\textsuperscript{6,12,13}.

- Some drugs listed are used under the cascade.

References


