Hepatic Portal Venous Hypoperfusion in Small Animals - Digestive System - Merck Veterinary Manual

Reduced hepatic portal venous perfusion causes characteristic histopathologic changes. The most common causes of portal venous hypoperfusion are congenital malformations impacting portal venous perfusion, including microvascular dysplasia and congenital portosystemic vascular anomalies.

Portal venous hypoperfusion (PVHP) is a histologic diagnosis distinguished by observation of close proximity of lobular elements (portal and centrilobular regions), small hepatocytes with an increased number of binucleated forms, increased frequency of miniaturized portal tracts, a dominating arterial perfusion typified by increased profiles of thick-walled muscular arterioles as well as serpiginous arterioles entangled with ductule profiles, and variable presence of portal vein silhouettes.

Diagnosis of portal hypoplasia cannot be made on the basis of a liver biopsy because any decrease in hepatic portal venous perfusion causes identical microanatomic change. This histologic pattern is better termed portal venous hypoperfusion.

Any disorder diminishing perfusion via the portal vein can lead to PVHP. Acquired disorders include abdominal mass lesions (eg, splenic mass compressing against the portal vein, other neoplastic lesions), chronic gastric torsion/dilation impacting portal venous circulation, fibrous entrapment of the portal vein, diaphragmatic herniation of a liver lobe, portal vein phlebitis (enteric vasculitis, severe inflammatory bowel disease, endotoxemia), portal vein thromboembolism, other intraluminal portal venous lesions [metastatic neoplasia]).

The most common causes of PVHP are congenital malformations impacting portal venous perfusion, including microvascular dysplasia (MVD) and congenital portosystemic vascular anomalies (PSVAs, also referred to as portosystemic shunts or portocaval shunts). Dogs are more commonly affected than cats.

The most common circulatory anomaly of the liver in dogs is microvascular dysplasia (MVD), shown to affect small-breed dogs with propensities for portosystemic vascular anomalies. MVD appears to share polygenic inheritance factors with PSVA in small breed terrier-type dogs. MVD is not recognized in cats or large-breed dogs. Commonly affected breeds, as well as mixes of

these breeds, include:

Yorkshire Terriers

Maltese

Bichon Frises

Cairn Terriers

Pugs

Miniature Schnauzers

Tibetan Spaniels

Norfolk Terriers

Scottish Terriers

Shetland Sheepdogs

Chihuahuas

Histologic features of MVD cannot distinguish this disorder from other causes of PVHP. Thus, liver biopsy is unrewarding to confirm diagnosis of MVD versus PSVA.

The predominant vascular malformation in MVD involves maldevelopment of tertiary portal vein branches as they arborize within the liver. This was initially detailed by examining radiographic portal venography, colorectal scintigraphy, and histologic features of different liver lobes in affected dogs. MVD differentially affects liver lobes, with the least affected being the caudate lobe perfused by the first branch of the portal vein.

Other microanatomic abnormalities observed in addition to classic findings of PVHP include:

increased numbers of thick-walled arterial cross-sections in portal tracts, serpiginous arterioles coiled around bile ductules, and "orphaned" random arterioles within the hepatic parenchyma

portal tracts fused with central veins (fusion complexes) sharing the same adventitial support

presence of unusual bifurcating portal vein structures external to the portal tract (abnormal sinusoidal inlet venules, so-called herniated portal veins)

increased numbers of small vascular structures in portal tracts that likely represent lymphatic vasculature and distended lymphatic surrounding hepatic venules—likely reflecting impact of arterialized sinusoidal perfusion driving increased ultralymph formation

random foci of disorganized distended hepatic sinusoids

prominent contraction of the spiral perivenular throttling muscle associated with hepatic veins

random distribution of small lipogranulomas laden with iron, reflecting small foci of hepatocyte degeneration

Dogs with MVD typically have no clinical signs (normal body size, no episodes of hepatic encephalopathy), no routine clinicopathologic test abnormalities as found in dogs with PSVA, no hyperammonemia, and no ammonium biurate crystalluria. MVD is usually recognized on discovery of increased total serum bile acid concentrations. While the fold increase in total serum bile acids is often less than encountered with PSVA, values cannot definitely differentiate these diagnoses.

Protein C activity is usually > 70%, contrasting with the common finding of low activity in dogs with PSVA. However, protein C as a stand-alone test cannot definitively differentiate a dog with MVD from a dog with PSVA.

Abdominal ultrasonography may disclose a subjectively small liver with decreased liver lobe portal vein perfusion. Colorectal scintigraphy confirms isotope transport to the liver before the heart (ruling out portosystemic shunting) and also may detail differential liver lobe perfusion.

Because MVD is characterized by increased total serum bile acid (TSBA) concentrations, this test should be administered to all puppies of predisposed breeds at 6 months old for future health-care considerations. This recommendation is made because discovery of high TSBA concentrations during an illness later in life may lead to inappropriate, invasive, and expensive diagnostic testing. Selection of breeding stock in affected kindreds should target dogs with normal TSBA concentrations. However, because the trait is polygenic, breeding dogs with normal TSBAs may still result in puppies affected with MVD and PSVAs.

Once high TSBA concentrations are detected in young (< 6 months) small terrier-type breeds lacking clinical signs of PSVA, repeated bile acid measurements are not warranted. In these dogs, TSBA concentrations will remain variably increased for the animal's life and will vacillate because of the physiologic variables influencing the bile acid test. Knowing that a dog has high bile acids likely due to MVD will define utility of the TSBA test for future health-care assessments.

Definitive diagnosis of MVD is possible only with liver biopsy combined with definitive vascular imaging studies. Liver biopsy demonstrates lesions identical to PSVA and other causes of portal venous hypoperfusion. Because the severity of MVD lesions varies among liver lobes, collection

of samples from three different lobes is recommended. Needle biopsy is strongly discouraged because recognition of PVHP is based on examination of multiple acinar units, which is restricted in small samples.

A normal lifespan should be expected in dogs with MVD. This diagnosis does not warrant feeding a special diet or liver-specific medications (eg, lactulose, SAMe, milk thistle, ursodeoxycholic acid, or other supportive hepatic measures). However, because dogs with MVD may have trouble metabolizing drugs that require rapid hepatic delivery and extraction, care is warranted when prescribing certain medications. For example, a sedation dose of butorphanol in MVD may be as low as 0.05 mg/kg, IV.

Dogs with vacillating liver enzyme activity (ALT predominantly) may have concurrent health issues. In most cases this involves inflammatory bowel disease (IBD) that has shared cytokines and inflammation causing focal hepatic injury. Some dogs with IBD and MVD display intermittent inappetence as the only sign of illness; this is particularly apparent in those with eosinophilic enteric infiltrates. Dogs with vacillating liver enzymes should receive bioavailable S-adenosylmethionine (SAMe; 20 mg/kg enteric coated tablets on an empty stomach) and low-dose vitamin E (10 U/kg, PO, every 24 hours with food) for antioxidant support.

Seemingly, dogs with MVD have increased risk for accumulating inflammatory mediators, cytokines, lymphocytes, plasma cells, and sometimes eosinophils in portal tracts and around central veins. In some, a degenerative centrilobular inflammatory lesion impacts transhepatic perfusion and can transform into a centrilobular hepatitis with a venoocclusive phenotype.

Responses to feeding of a hypoallergenic diet combined with low-dose metronidazole (7.5 mg/kg, PO, every 12 hours) and exclusion of endoparasitism are used as a clinical trial before pursuit of tissue biopsy. Liver and intestinal biopsies are needed to characterize the hepatic injury pattern and IBD if the initial clinical trial fails to resolve liver enzyme activity. Biopsy is done to confirm a diagnosis and determine whether immunomodulation is warranted. Dogs with eosinophilic inflammation resistant to feeding of a hypoallergenic diet with concurrent low-dose metronidazole require glucocorticoids at a minimum.

A portosystemic vascular anomaly (PSVA) is a congenital, grossly apparent aberrant connection between the extrahepatic portal vasculature and the systemic circulation (connecting a branch of the portal vein to the vena cava or azygous vein) that diverts blood to the systemic circulation, bypassing the liver. Diminished portal flow to the liver leads to hepatic lobular atrophy due to diversion of essential hepatotrophic substances carried in the splanchnic circulation (largely from the pancreas).

Because the portal circulation transports microorganisms, toxins, nutrients, and other materials from the intestines to the liver, detoured blood is not cleansed or processed before systemic distribution. Consequently, neurotoxic substances that can provoke hepatic encephalopathy (HE) are circulated directly to the brain. Other noxious products and infectious agents inefficiently removed by the liver in animals with PSVAs result in a more severe clinical challenge to what should be mild health concerns (eg, infectious diarrhea, infected wounds, dermatitis, and tick bites).

While PSVAs are congenital, extensive genetic mapping studies have failed to define a single genetic defect, although certain breeds are predisposed to extrahepatic portosystemic vascular anomalies (E-PSVAs) and certain breeds to intrahepatic portosystemic vascular anomalies (I-PSVAs). E-PSVAs predominantly affect small-breed dogs and cats whereas I-PSVAs predominantly affect large-breed dogs. However, there are many exceptions.

E-PSVAs are predominantly encountered in small purebred terrier-type dogs (eg, Yorkshire Terriers, Maltese, Shih Tzus, Havanese, Papillons, Miniature Schnauzers, Pugs, Cairn Terriers, Norfolk Terriers, Tibetan Spaniels) and mixes of these breeds. Intrahepatic PSVAs predominate in large-breed dogs, including (but not exclusively) Labrador Retrievers, Golden Retrievers, Irish Wolfhounds (kindred related inheritance shown), and Old English Sheepdogs.

The most common types of E-PSVAs involve shunting via normal portal tributaries rather than via anomalous vasculature. Based on numerous retrospective clinical studies, the most common E-PSVAs in dogs (> 90% of cases) include splenocaval, left gastrophrenic, left gastroazygous, and numerous additional shunts involving the right gastric vein. In cats, > 90% of E-PSVAs include splenocaval, left gastrophrenic, and left gastrocaval, with fewer left gastroazygous. However, some specific shunts described in the veterinary literature have been erroneously characterized. Detailed studies by one group proposes that regardless of shunt type, abnormal communications usually involve the left gastric vein.

An intrahepatic PSVA represents retention of the ductus venosus, an embryonic connection between the umbilical vein and caudal vena cava that carries fetal blood from the placenta to the heart, through the liver, bypassing the hepatic circulation. This malformation is only occassionally seen in small-breed dogs and cats. I-PSVAs are traditionally described according to their position in the liver as left, central, or right divisional shunts. Morphology of ductus venosus in neonatal puppies is identical and consistent with morphology of left divisional I-PSVAs; consequently, these can be termed patent ductus venosus. It remains unclear if central or right divisional I-PSVAs are aberrant patent ductus venosus or anomalous vasculature.

Congenital PSVAs in cats are encountered more frequently in mixed breeds; however,

prevalence may be increased in purebred Himalayans and Persians. The higher prevalence of polycystic liver disease and associated portal hypertension and acquired portosystemic shunts in these breeds complicates diagnosis of PSVAs. In cats, extrahepatic PSVAs involving the left gastric vein are most common.

Animals with PSVAs are often smaller than littermates, fail to thrive, and can have other congenital abnormalities (eg, cryptorchidism in dogs and cats, heart murmurs in cats, rare cardiac malformations, ductal plate malformations). Clinical signs are highly variable with 10%–20% of affected animals clinically normal. Clinical signs depend on the severity of portosystemic shunting and, in clinically affected animals, are usually evident by 6 months old in cats and before 1 year in dogs.

Clinical signs of PSVAs include nausea, ptyalism (especially cats), vomiting, diarrhea, pica, intermittent anorexia, polyuria/polydipsia (PU/PD), amaurosis (unexplained blindness), excessive vocalization, hallucinations, apparent neck or spinal pain, apparent transverse myelopathy, signs of polyarthritic pain, hematuria, pollakiuria, stranguria, urethral obstruction associated with formation of ammonia biurate uroliths, and additional neurobehavioral signs reflecting HE.

Clinical signs referable to urinary tract calculi may be the only presenting complaint. Cats with PSVAs have a unique, homogenous, copper-colored iris that appears to be genetically linked with the disorder; the exception is in blue-eyed cats. Because a copper-colored iris is typical for Persians and Russian blue cats that do not have a PSVA, it is important to coordinate this normal variation with clinical signs and finding high TSBA concentrations.

Laboratory abnormalities in animals with PSVAs may include microcytic RBCs (low mean corpuscular volume), mild nonregenerative anemia based on PCV but with normal RBC count, poikilocytosis (cats), target cells (dogs), mild hypoproteinemia and hypoalbuminemia, hypoglycemia (rare, primarily young, very small toy-breed dogs), low BUN and creatinine, hypocholesterolemia, normal to mildly increased liver enzyme activity (ALT, AST, and alkaline phosphatase [ALP], ALP reflecting juvenile bone growth), normal bilirubin, dilute urine (hyposthenuria or isosthenuria), and ammonia biurate crystalluria.

Fasting and postprandial TSBA concentrations are usually markedly increased (> 150 uM/L); however, measurement of TSBAs or ammonia after a prolonged fast may yield normal values. Postprandial TSBAs and ammonia (after NH_4Cl administration) are markedly abnormal.

Routine coagulation assessments are usually within normal limits; however, protein C activity is usually < 70%. The protein C test is valid for use in dogs but not in cats. This test reflects the

severity of shunting; the lower the value, seemingly the more severe the shunting. In clinically normal PSVA dogs with a protein C > 70%, surgical ligation has been predictably successful. In dogs with a protein C < 70%, some do tolerate complete shunt attenuation.

Abdominal radiographs of patients with PSVAs disclose microhepatia and plump kidneys. Because ammonium biurate uroliths are radiolucent, these are not detected radiographically unless chronic urinary tract infection has generated a mixed struvite-urate concretion. Ultrasonography can noninvasively identify PSVA if the study is completed by an experienced operator using color-flow Doppler in a fasted, cooperative patient. Typical findings include subjective identification of microhepatia, plump kidneys, and renal or urinary bladder uroliths.

Specific ultrasonographic identification of E-PSVAs can be challenging because of interference by bowel gas and lack of patient cooperation that may limit imaging in critical regions. E-PSVAs can be missed with ultrasonography and even falsely identified in animals when a convincing history has biased the imaging operator. Ultrasound confirmation of I-PSVA is comparatively easy.

In animals with PSVAs, measurement of the cross-sectional diameter of the portal vein (PV) and aorta (Ao) at the level of portal hepatis is used to calculate a PV:Ao ratio. Finding a PV:Ao < 0.65 is consistent with a PSVA or extrahepatic portal atresia. The PV is measured just before entrance into the liver. As the Ao fluctuates during the cardiac cycle, video frames are used to capture its maximum diameter for the PV:Ao ratio.

The PV:Ao ratio is used to correct for differences in body size among dogs but also be applied to cats. Discovery of vena caval distension and turbulent blood flow at or just caudal to this site (ie, random chaotic multidirectional high-velocity flow) usually identifies the union between a PSVA and vena cava.

Colorectal portal scintigraphy or splenoportal scintigraphy, available in specialty clinics or teaching hospitals, can define presence or absence of portosystemic shunting. However, scintigraphy is unable to definitively identify the anatomic location of shunting vasculature. Colorectal scintigraphy is simply done by deep rectal injection of an isotope-laden enema. Splenoportal scintigraphy requires percutaneous injection of isotope into the spleen and allows smaller isotope dosing but does not have greater clinical utility for detection of portosystemic shunting compared to the colorectal procedure.

Contrast radiographic portography, the historic gold standard for confirming PSVAs, requires catheterization of a mesenteric portal vein branch and injection of radiodense (iodinated) contrast. Image interpretation is usually straightforward. Currently, this imaging modality is

used as an intraoperative fluoroangiographic procedure during shunt attenuation in hospitals using a direct ligation method. After identification of the suspected PSVA, the shunt vessel is marked with umbilical tape, and a portogram is completed to confirm correct vessel identification.

The surgeon strategizes the best site for shunt attenuation (as close as possible to the site of anastomosis with the vena cava) and a temporary ligation is applied, monitoring portal pressure change and physiologic response to shunt attenuation (heart rate, blood pressure, gross perfusion of intestines, and their motility). A second portogram is used to illustrate the impact of shunt attenuation on the intrahepatic portal perfusion. If ligation appears effective, it is tightened to tolerance (may be complete or incomplete attenuation) for permanent attenuation with silk.

Noninvasive multisector CT has replaced preoperative contrast radiographic portography for definitive diagnosis of PSVA in most hospitals. This imaging modality requires short-term general anesthesia with contrast injected into a peripheral vein. Image capture allows three-dimensional reconstruction of splanchnic portal, vena caval, and arterial vasculature and also details presence of renal or urinary bladder urolithiasis. Image interpretation requires an experienced clinician who can accurately identify splanchnic vasculature and deduce the site of PSVA and vena caval anastomosis.

Typically, the lumen of the vena cava expands in width at the site of PSVA anastomosis.

This methodology allows detection of portoazygous and portosplenophrenic shunting vasculature, which are sometimes difficult to identify. Coordination of image capture with venous and arterial contrast phases increases interpretation accuracy.

It is possible that these patients have perfusion of shunting vasculature influenced by the gravitational effects of surrounding viscera or postural influences that minimize shunt flux in certain positions (ie, less shunting when standing). This consideration is predicated on the inability to image some PSVAs with portovenography in certain recumbent (lateral) postures. Surgical management of E-PSVA may include careful direct intraoperative graded shunt ligation at surgery to the animal's tolerance (judged by surgeon assessment of measured portal pressure [water manometer], systemic blood pressure, heart rate, and local visceral response) or use of methods believed to achieve gradual shunt attenuation.

These latter methods include application of an ameroid constrictor (metal ring with an inner absorptive lining that gradually expands with fluid absorption over a few days, slowly occluding shunting vasculature) or placement of a cellophane band around the PSVA thought to gradually

initiate an occlusive inflammatory response. There is no consensus regarding the best surgical method for PSVA management.

A recent meta-analysis of published case series concluded better outcomes occur with direct complete shunt ligation or ameroid banding versus placement of a cellophane band or incomplete ligation. However, a major obstacle confounding assessment of various surgical interventions is the naive perception that slow shunt closure can be accommodated by all animals and is safer than intrasurgical ligation, discounting the wide variability in extent of microvascular hepatic malformations among these patients. Some patients have various degrees of portal vascular atresia, which precludes full shunt attenuation by any method.

Surgical intervention for intrahepatic shunts is more complicated and less successful. Left-sided I-PSVAs are sometimes amenable to intraoperative ligation.

I-PSVA attenuation by endovascular thrombotic coiling by interventional radiographic technique is an alternative. This can be associated with adverse effects (portal hypertension, portal or vena caval clot formation) and can be prohibitively expensive; more than a single intervention may be needed to substantially curtail the portosystemic shunting. This procedure involves fluoroscopic placement of a vascular stent in the vena cava, across the site of anastomosis with the I-PSVA.

Subsequently, a vascular catheter is inserted across the stent wall into the shunt lumen where thrombotic coils are carefully deployed. Sequential assessment of intravascular shunt pressure guides procedural endpoint. Gradual increase in shunt attenuation occurs over the following weeks.

Coexistent DPM leads to splanchnic hypertension and APSSs as a result of presinusoidal PH associated with severe small proliferative DPM phenotypes (classified as congenital hepatic fibrosis). In these, PSVA attenuation eliminates decompression of the splanchnic circulation, allowing the impact of the presinusoidal fibrosis to manifest. Liver biopsies should be collected at the time of PSVA ligation from three liver lobes, avoiding sampling of the caudate lobe, which receives the first intrahepatic portal vein branch and demonstrates the fewest features of portal venous hypoperfusion.

Observation of severe liver lobe atrophy or atresia, absence of a gallbladder, or observation of an atretic gallbladder are features commonly associated with DPM. Discovery of APSSs at surgery in a young dog or cat rules out diagnosis of PSVA, which would decompress the splanchnic bed and indicate greater likelihood of a congenital hepatic fibrosis DPM phenotype. These vessels should not be attenuated.

Male dogs with repeated bouts of ammonium biurate urolithiasis, despite medical or surgical management of PSVA-related hyperammonemia, should have a permanent prescrotal urethrostomy created to allow passage of small calculi. Ammonium urate uroliths do not dissolve with any medical intervention or diet change. Thus, cystic calculi should be removed at the time of urethrostomy. Adjusted medical management (usually dietary modifications) along with urethrostomy should avert future occurrence of an obstructive uropathy. Allopurinol is not a recommended intervention for these patients.

In dogs, the most common postsurgical complication of E-PSVA attenuation is short-term benign abdominal effusion that typically resolves within a few days. The most serious postsurgical complication is **acute portal hypertension**, characterized by abrupt development of abdominal effusion, bloody diarrhea, abdominal pain, ileus, endotoxic shock, and cardiovascular collapse. This complication requires immediate release of shunt occlusion but is often fatal.

Other complications include seizures (rare) and formation of portal venous thrombi. Another serious postsurgical complication is development of intractable recurrent seizure activity within 7 days of shunt ligation. Postattenuation seizures affected ~5%–8% of dogs; however, published incidence is widely variable among institutions and hospitals. A multi-institutional retrospective study involving 940 dogs undergoing shunt attenuation demonstrated no protective benefit from administration of levetiracetam as a presurgical treatment.

Postattenuation seizures are far more common as a severe complication in cats, estimated to affect ~20%–30% of patients with incidence as high as 75% in small case series. A subset of dogs and cats unobtrusively develop APSSs over weeks, months, or years after E-PSVA or I-PSVA attenuation. Development of APSSs is more common in cats than dogs; partial ligation with staged surgeries to achieve complete ligation in cats has not abrogated this complication. The site of shunt ligation also may transform into a medusa or variceal of shunting vessels, or the ligated shunt may recanalize, reestablishing portosystemic shunting several years after an attenuation procedure.

The greatest risk for insidious postoperative complication occurs with ameroid constrictors and cellophane banding because the extent of vessel occlusion remains ill defined unless repeat imaging studies are undertaken. Ameroid constrictors also may be complicated by unexpected accelerated shunt attenuation (device twisting, premature closure) as well as shunt attenuation beyond patient tolerance. The latter problem results in development of portal hypertension and APSSs. An additional complication with ameroid devices is that they may fail to achieve intended complete PSVA attenuation in animals that could have tolerated complete shunt

occlusion at the time of surgical intervention.

Lastly, in rare cases, ameroids have eroded through shunting vasculature, causing acute collapse, hemoabdomen, and death, months to years after their application. Cellophane band failures are well documented and relate either to failure of proper perivascular placement or to the type of cellophane product used; some products fail to initiate the expected inflammatory response. When ameroid or cellophane banding is used, thoughtful intraoperative assessment of patient response to full shunt attenuation is advised because there is no assurance that slow attenuation to complete occlusion will be tolerated without development of APSSs.

Physical findings: After successful PSVA attenuation, liver size should increase and clinical signs, including PU/PD and episodic neuroencephalopathic features, should abate. The copper-colored irises in cats with PSVA do not change; this is considered a gene-linked phenomenon.

Routine hemogram, serum biochemistry: Successful shunt attenuation is associated with resolution of microcytosis; normalization of BUN, creatinine, and cholesterol concentrations, and disappearance of dilute urine concentration and ammonium biurate crystalluria.

Total serum bile acids: With successful shunt attenuation in small-breed dogs, it is common for TSBAs to decline but remain abnormal, reflecting either partial PSVA closure or the common affiliation of MVD. In large-breed dogs, this directly reflects incomplete shunt attenuation.

Protein C activity: In dogs with presurgical subnormal protein C activity (< 70%), repeated assessment after 2 months allows better prediction of the success of shunt attenuation than is possible with measurement of TSBA concentrations in small-breed dogs. In dogs achieving a clinically relevant decrease in their shunt fraction, protein C will normalize. However, this does not assure complete shunt attenuation but rather documents notable clinical benefit of shunt attenuation.

There is no consensus regarding the best imaging procedures for postoperative assessment of the completeness of shunt attenuation and gain in hepatic portal perfusion. Ultrasound evaluation is subjective and image interrogation complicated by enteric contents (food, gas) and patient cooperation. Colorectal scintigraphy determines only macroscopic shunting. Spiral CT or MRI requires general anesthesia; CT can be complicated by asynchrony between contrast injection and vascular phase capture and poor discrimination of intrahepatic vascular detail. These studies also can be difficult to interpret.

Splenoportal portography (injection of iodinated contrast into the splenic parenchyma or splenic vein for radiographic portography) does not deliver enough contrast to definitively define shunting vasculature and details of hepatic perfusion. Full mesenteric portography requires

general anesthesia and is too invasive for routine follow-up assessments. Minimally invasive fluoroscopic angiography (femoral artery to hepatic artery) requires image capture during the second-pass venous phase and lacks needed detail.

Survival analyses of > 450 dogs with PSVAs treated by surgical shunt attenuation versus primary longterm medical management without surgical intervention in the author's hospital (98% of surgeries completed by two board-certified surgeons) described a median survival time of 2,555 days in dogs with E-PSVAs treated surgically (n = 195) and 2,109 days for dogs treated medically (n = 169); survivals in this population were not significantly different. Results are biased because owners of dogs optimally responding to medical management were inclined to decline surgical intervention. Furthermore, at least 50% of dogs with surgical management continued with some form of medical intervention.

Findings demonstrate efficacy for medical management in some dogs if financial constraints limit surgical options or if patient response is astoundingly positive. Conversely, reports of 124 dogs managed at four European referral hospitals where cases were prospectively entered by owner into medical (n = 27) or surgical (n = 97) interventions (direct ligation [39 complete, 24 partial], ameroid ligation [29], cellophane banding [5]) had multiple surgeons involved and reported dismal survival for medically managed dogs (median survival of 827 days medically managed compared to 2,156 days for surgically managed dogs). Quality of life assessments for dogs on medical management were lower than for surgically corrected dogs.

Median survival time in dogs with I-PSVAs treated surgically in the author's hospital (n = 46), again with a single primary surgeon, was 1,215 days and not significantly different from dogs treated by medical management alone (n = 48) of 1,423 days. Longer survival times for dogs with I-PSVA have been reported for dogs undergoing minimally invasive endovascular coil embolization (in one report of 96 dogs with I-PSVA treated by one or more coil embolization procedures by a single interventional group, median survival time was 2,204 days).

Overall, prognosis after surgical ligation of a single E-PSVA in dogs is usually good but is less favorable in dogs that develop APSSs. Development of APSSs signals presence of severe intrahepatic portal vein atresia or ductal plate malformation with a congenital hepatic fibrosis phenotype. Prognosis for dogs with I-PSVA with successful surgical attenuation or transvenous embolic coiling also can be fair to good. Transvenous embolic coiling is usually more expensive than surgical intervention of E-PSVA.

Medical management to avoid development of hepatic encephalopathy and ammonium biurate crystalluria is a primary objective in all patients with PSVAs. Before scheduling surgical intervention, balanced medical support should be instituted to optimize patient neurologic

status. Generally, this is done for at least a week to improve patient tolerance to anesthetics.

Young puppies of toy breed dogs with tiny stature on first presentation are supported with medical management while they grow to make them better candidates for imaging studies and manual shunt manipulation. Medical management remains an option for chronic patient care in minimally clinically affected animals.

Good quality of life and normal life span can be achieved in those demonstrating strong positive response to medical interventions. However, patients managed medically remain at risk for dramatically severe illnesses escalating from what would normally be considered benign problems. Of particular importance are infections and gastrointestinal disorders increasing risk for translocation of enteric toxins and bacteria. Decline in Kupffer cell surveillance due to hepatofugal portal circulation (bypassing this important population of fixed macrophages) allows even minor infections to escalate into serious systemic illness.

Nutritional support is the cornerstone of medical care for patients with PSVA. For dogs, prescription diets formulated for management of canine hepatic insufficiency are recommended. Lifelong dietary support will be required in patients that do not undergo PSVA attenuation, cannot tolerate full shunt ligation, or fail to improve after PSVA attenuation with an ameroid or cellophane banding.

Slow incremental titration of additional protein supplements at 0.25 g protein/kg for 5–7 day intervals is monitored for safety. The longterm objective is to achieve an additional 1–1.5 g protein/kg intake. Shredded cheese, cottage cheese, and yogurt are often used as convenient sources. Treats of raw vegetables (eg, broccoli, carrots), cheese, probiotic yogurt, popcorn, cooked squash, certain breakfast cereals without artificial sweeteners, and human animal crackers are safe treats.

Occasional dog biscuits and limited supervised activity with rawhide bones (US manufactured) can be allowed without adverse consequences. If rawhide chews result in oral hemorrhage, they should be avoided because swallowed blood can provoke encephalopathic signs.

In addition to dietary intervention, administration of lactulose and low-dose metronidazole is advised. Lactulose dosing must be titrated to achieve several soft puddinglike stools per day, avoiding overdosage, which can lead to painful cramping (fermentation gas) and severe diarrhea. Many dogs and cats appreciate the taste of lactulose such that it may be given independently or mixed with food.

Metronidazole dosing must not exceed 7.5 mg/kg, PO, every 12 hours, else neurologic signs may develop (ataxia, seizure activity). Neomycin is not recommended because of risks for ototoxicity

(deafness) and renal injury that have been observed in PSVA patients treated chronically with this medication.

It is important to warn the owner that medically managed patients without PSVA attenuation remain at risk for developing HE. Owners should be educated to recognize early signs of HE and complicating health issues, to administer cleansing and retention enemas, and to administer subcutaneous fluids. These early interventions can minimize HE episodes and decrease costly emergency veterinary visits that may lead to patient euthanasia.