

Congenital Portosystemic Shunt or Vascular Anomaly

Basics

OVERVIEW

- Congenital (present at birth) portosystemic shunt or portosystemic vascular anomaly—malformation of the veins connecting the portal and general body (systemic) circulations, permitting portal blood to bypass the liver; the portal vein is the vein that normally carries blood from the digestive organs to the liver
- Malformation of the veins may be within the liver (known as an “intrahepatic shunt”), more common in large-breed dogs, or outside the liver (known as an “extrahepatic shunt”), more common in small-breed dogs and cats
- Most shunts are single blood vessels
- Many small-breed dogs also have other blood vessel (vascular) abnormalities involving the small blood vessels within the liver (known as “intrahepatic microvasculature”)
- Acquired (condition that develops sometime later in life/after birth) portosystemic shunt (condition of abnormal blood flow in the liver due to high blood pressure in the portal vein [known as “portal hypertension”], the vein carrying blood from the digestive organs to the liver)—can develop following surgical “tying off” or “ligating” of the congenital (present at birth) abnormal blood vessel
- The liver is the largest gland in the body; it has many functions, including production of bile (a fluid substance involved in digestion of fats); production of albumin (a protein in the plasma of the blood); and detoxification of drugs and other chemicals (such as ammonia) in the body



GENETICS

- Genetically transmitted in high-risk breeds
- Affected breeds—Yorkshire terriers, cairn terriers, Maltese, Tibetan spaniels, miniature schnauzers, Norfolk terriers, pugs, shih tzus, Havanese, Irish wolfhounds, Old English sheepdogs, and others
- Suspect inheritance as a dominant trait, with incomplete penetrance

SIGNALMENT/DESCRIPTION OF PET

Species

- Dogs (more common)
- Cats

Breed Predispositions

- Higher risk—purebred dogs; mixed-breed dogs of small terrier descent; mixed-breed cats

- Yorkshire terriers, Maltese, cairn terriers, Tibetan spaniels, miniature schnauzers, Norfolk terriers, pugs, shih tzus, Havanese, Irish wolfhounds, Old English sheepdogs

Mean Age and Range

- Usually first identified in juvenile pets; but dogs have been as old as 13 years of age at first diagnosis
- Accidental discovery of presence of portosystemic shunt: older pets that do not have clinical signs

SIGNS/OBSERVED CHANGES IN THE PET

- Pet may have normal appearance or a stunted stature; stunted growth—common
- Signs often initiate with weaning of puppy or kitten to commercial food
- Gastrointestinal signs—lack of appetite; vomiting; diarrhea; eating of nonfood items (known as “pica”)
- Cats initially thought to have upper respiratory infection (due to display of excessive drooling [known as “ptyalism”], which is a sign of hepatic encephalopathy in cats)
- Episodic brain disorder caused by accumulation of ammonia in the system due to inability of the liver to rid the body of ammonia (known as “hepatic encephalopathy”)—episodes transiently improve with fluid therapy, broad-spectrum antibiotics, and lactulose
- Central nervous system signs—weakness; pacing; wobbly, incoordinated or “drunken” appearing gait (known as “ataxia”); disorientation; head pressing; blindness; behavioral changes: aggression (cats), vocalization, hallucinations; seizures; coma
- Urinary signs—increased urination (known as “polyuria”) and increased thirst (known as “polydipsia”); presence of ammonium biurate crystals in the urine; abnormal frequent passage of urine (known as “pollakiuria”); difficult or painful urination (known as “dysuria”); blood in the urine (known as “hematuria”); blockage or obstruction of the urethra (the tube from the bladder to the outside, through which urine flows out of the body) and rarely the ureters (the tubes from the kidneys to the bladder) due to the presence of ammonium biurate urinary tract stones (known as “ammonium biurate uroliths”)
- Some dogs (approximately 20% of affected pets) lack clinical signs
- Affected female dogs (bitches) may produce litters
- Affected dogs may be used at stud before diagnosis recognized
- Small liver (known as “microhepatica”)
- Copper-colored irises in non-blue-eyed cats (note: Persian, Russian blue, and some other cats normally have copper-colored irises)
- Fluid buildup in the abdomen (known as “ascites”) or in other tissues of the body (known as “edema”)—rare

CAUSES

- Congenital (present at birth) malformation of blood vessels
- Acquired (condition that develops sometime later in life/after birth) portosystemic shunt in pets with congenital (present at birth) portosystemic vascular anomaly may develop subsequent to increased blood pressure in the portal vein (portal hypertension), either congenital portal hypertension or surgically induced portal hypertension following surgical “tying off” or “ligating” of the abnormal blood vessel

RISK FACTORS

- Portosystemic shunt or vascular anomaly—purebred dogs, especially small terrier-type breeds
- The Irish wolfhound appears to have slow closure of the fetal blood vessel (known as the “ductus venosus”) that carries blood from the umbilical vein to the vena cava; the “vena cava” is the main vein that returns blood from the body to the heart; early screening may incorrectly identify dogs as being affected with portosystemic shunt or vascular anomaly

Treatment

HEALTH CARE

- Inpatient—severe signs of hepatic encephalopathy (brain disorder caused by accumulation of ammonia in the system due to inability of the liver to rid the body of ammonia); supportive care and initiation of medical management prior to liver biopsy and surgical ligation
- Hepatic encephalopathy should be treated medically before surgery to “tie off” or “ligate” blood vessels

DIET

- Nutritional support—essential to maintain body condition, as muscle serves as an important site of temporary ammonia detoxification
- Balanced, protein-restricted diet—recommended; thereafter, protein allocation based on response in combination with treatment for hepatic encephalopathy; as tolerated, add protein (use cottage cheese or calcium caseinate in dogs), as directed by your pet's veterinarian

SURGERY

- Congenital (present at birth) portosystemic shunt or portosystemic vascular anomaly (in which blood flows abnormally between the portal vein [vein that normally carries blood from the digestive organs to the liver] and the body circulation without first going through the liver)—surgical correction (in which the abnormal blood vessel is “tied off” or “ligated” using Ameroid constrictor or cellophane banding) is a possibility in many cases
- Surgical ligation—optimal goal is total ligation, but this may not be tolerated in some dogs
- Partial ligation only may be achieved in many dogs and generally leads to improved health
- Ameroid constrictor—reduces immediate surgical risks of ligation; may later result in acquired (condition that develops sometime later in life/after birth) portosystemic shunt in some pets (especially Yorkshire terriers)
- Portosystemic vascular anomaly within the liver (intrahepatic)—most difficult to ligate
- Emergency surgery—sometimes required for removal of ligature or Ameroid constrictor
- Fluid buildup in the abdomen (ascites)—common after shunt ligation; may be sign of increasing blood pressure in the portal vein (portal hypertension)
- Intensive care (ICU) monitoring—recommended post-operatively for 72–96 hours
- Anesthesia and surgery do carry risks—5–25% mortality rate, depending on various factors including type of portosystemic shunt or vascular anomaly, other liver abnormalities, and supportive critical care

Medications

Medications presented in this section are intended to provide general information about possible treatment. The treatment for a particular condition may evolve as medical advances are made; therefore, the medications should not be considered as all inclusive

- Medical management is directed at treatment of hepatic encephalopathy (brain disorder caused by accumulation of ammonia in the system due to inability of the liver to rid the body of ammonia)
- Medications that increase dietary protein tolerance, change bacteria or conditions in the intestines, or reduce production or availability of substances provoking hepatic encephalopathy
- Antibiotics—antibiotic selection based on ability to change the bacteria in the intestines or their products; administered by injection (known as “systemic administration”); antibiotics such as metronidazole or amoxicillin; combine use with lactulose
- Nonabsorbable-fermented carbohydrates—lactulose, lactitol, or lactose (if lactase deficient); decrease production or absorption of ammonia; increase rate of stool transit; trap nitrogen in bacteria; lactulose most commonly used; therapeutic goal is passage of two to three soft stools daily; also may be administered as an enema for sudden (acute) hepatic encephalopathy and coma *after* cleansing enemas have removed debris
- Probiotics with nonabsorbable-fermented carbohydrates—may be helpful in changing the intestinal bacteria to decrease production of toxins that contribute to hepatic encephalopathy
- Enemas—*cleansing enemas* (warmed polyionic fluids) mechanically clean colon; *retention enemas* directly deliver fermentable substrates or directly alter colonic pH and organisms: diluted lactulose, lactitol, or lactose; neomycin in water; diluted Betadine
- Zinc supplementation, as directed by your pet's veterinarian
- Fluid buildup in the brain (known as “cerebral edema”)—complicates sudden (acute) hepatic encephalopathy (brain disorder caused by accumulation of ammonia in the system due to inability of the liver to rid the body of ammonia); administer medication (mannitol) to decrease fluid buildup; administer nasal oxygen and N-acetylcysteine; use of steroids to decrease fluid buildup (edema) is controversial as steroids may promote bleeding in the intestinal tract (which is a risk factor for development of hepatic encephalopathy)
- If epileptic seizure activity—levetiracetam is the preferred medication to control seizures (known as “anticonvulsants”); other medications include zonisamide and potassium bromide

Follow-Up Care

PATIENT MONITORING

- Following surgical ligation of blood vessels, watch closely for signs of lack of blood flow to the intestines (such as bloody diarrhea, abdominal pain, failure to recover from surgery/anesthesia, unexplained rapid heart rate [known as “tachycardia”], increased body temperature [known as “hyperthermia”] or decreased body temperature [known as “hypothermia”]); monitor girth and body weight
- Reevaluate the pet's at-home behavior; body condition; girth circumference
- Monitor bloodwork (complete blood count [CBC] and serum biochemistry panel), and urinalysis (looking for resolution of ammonium biurate crystals in the urine [crystalluria])
- Monitor protein C—presurgical low protein C increases or returns to normal levels following successful surgery (ligation); protein C is a substance in the body that prevents blood from clotting—it is dependent on vitamin K, which is produced by the liver; liver disease can lead to low protein C levels

PREVENTIONS AND AVOIDANCE

- If multiple abnormal blood vessels (portosystemic shunts) are identified by ultrasound, they likely are acquired (condition that develops sometime later in life/after birth) portosystemic shunts—do not pursue surgical ligation; another underlying liver disease or disorder is causing increased blood pressure in the portal vein (portal hypertension)

POSSIBLE COMPLICATIONS

- Post-operative complications—blood clots in the portal vein (known as “portal venous thrombi”); sudden (acute) severe high blood pressure in the portal vein (portal hypertension); lack of blood flow to the intestines; accumulation of bacterial toxins in the blood (known as “endotoxemia”); seizures; generalized bacterial infection (known as “sepsis”); sudden (acute) inflammation of the pancreas (known as “pancreatitis”); bleeding
- Low body temperature (hypothermia) during or following surgery—especially in very small pets; complicates recovery
- Seizures following surgical ligation

EXPECTED COURSE AND PROGNOSIS

- Cannot predict individual response to surgery
- Dogs—surgical ligation improves signs in 70–80% of pets with clinical signs of portosystemic shunt
- Cats—many develop acquired (condition that develops sometime later in life/after birth) portosystemic shunt
- Following surgery—continue management of hepatic encephalopathy (brain disorder caused by accumulation of ammonia in the system due to inability of the liver to rid the body of ammonia) until reevaluation of clinical status
- Some pets require indefinite treatment
- Increased risk for poor surgical outcome in certain small dogs and cats
- Despite initial good response, recurrence of shunting may develop after 3 years
- Dogs with portosystemic vascular anomaly that do not have clinical signs and have not had surgery can have a full life expectancy

Key Points

- Discuss all treatment options (medical and surgical) with your pet's veterinarian
- Surgical ligation—expect improvement but not cure; may not be required for all dogs, as some respond well to feeding a commercial diet manufactured for pets with hepatic encephalopathy (brain disorder caused by accumulation of ammonia in the system due to inability of the liver to rid the body of ammonia)
- Clinical signs may persist despite surgical intervention (ligation), requiring long-term (chronic) nutritional and medical management
- If surgery is not performed or ligation is not tolerated, watch the pet carefully for signs of urinary tract problems, which might indicate the presence of ammonium biurate urinary tract stones (ammonium biurate uroliths); take your pet to the veterinarian if you observe any abnormal urinary tract signs