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The Berner Owner's Guide to Intrahepatic Portosystemic Shunts (IHPSS) or Liver Shunts

Berner Owner's Guide Series, Guide # 1
By Nancy P. Melone, Ph.D.

What is IHPSS?

All puppy fetuses have a large shunt (ductus venosus) that carries blood rapidly through the fetal liver to the heart. While the puppy is in the womb, the mother's liver does the work of filtering out toxins, storing sugar and producing protein for the unborn puppy. The puppy's liver does not function, yet. Within a week after birth, the shunt normally closes and the puppy's own liver begins to take over the tasks previously handled by the mother's liver. In puppies with liver shunts, the shunt never closes (*patent ductus venosus*), resulting in blood recirculating back through the brain and body, unfiltered of toxins.

What are the Symptoms of IHPSS?

Symptoms are often seen at a young age (under 1 year old). Some veterinarians believe that a number of undiagnosed "fader" puppies may suffer from IHPSS. Symptoms include intermittent anorexia, small stature (*i.e.*, puppy is often born normal size, but fails to grow after birth), poor muscle development, neurologic dysfunction (*e.g.*, circling, disorientation, unresponsiveness, staring, head pressing particularly after eating), unusual vocalization, seizures, and lethargy. Less frequent symptoms include excessive water consumption and frequent urination, urinary stones, diarrhea, vomiting, pica, or apparent blindness. Some dogs are diagnosed after they take a long time to recover from anesthesia or sedatives. Sometimes an IHPSS-affected dog will show no symptoms until the dog is older.

How is IHPSS Diagnosed?

Typically, diagnosis is done in three stages. First, your veterinarian will do a complete blood count and serum chemistry profile. Dogs with IHPSS typically have low blood urea nitrogen (BUN) and albumin concentrations. They may also be anemic or suffer from microcytosis (the dog's red blood cells are smaller than normal). Liver enzymes (AST and ALT) may be high. If a urinalysis is done, findings may include low specific gravity (dilute urine), ammonium biurate crystals, and abnormal pH.

After reviewing these findings, if your veterinarian suspects IHPSS, she will likely suggest running a fasting (pre-prandial) and a post-feeding (post-prandial) bile acid test. Your dog will be required to fast for 12 hours before the pre-prandial blood sample is drawn. He will then eat a meal and in 2 hours the post-prandial blood sample will be drawn. Dogs with IHPSS typically have elevated bile acid concentrations.

Unfortunately, IHPSS cannot be diagnosed definitively by blood work. A liver shunt can only be found using techniques such as ultrasound, scintigraphy, portography, CT, MRI or exploratory surgery. Each technique has advantages and disadvantages.

What are IHPSS Treatment Options?

There are two basic treatment alternatives -- medical management or surgery.

Medical Management

The least invasive alternative to stabilize the IHPSS-affected dog is to manage him medically. Typically this involves administering medications, such as lactulose, several times a day to reduce the amount of toxins produced and absorbed in the large intestine. In addition, the dog is usually put on a prescription low-protein/low-fat diet. If clinical signs are still not managed, then antibiotics (*e.g.*, metronidazole) are given to reduce the number of toxin-producing bacteria in the intestine. Other medications may be required to control seizures, blood sugar or to remove intestinal toxins. Most dogs see clinical improvement with medical management, but unfortunately over 50% of these dogs are euthanized within 10 months of their diagnosis because of uncontrolled seizures.

Traditional Surgery

According to most veterinarians, surgery provides the best chance for a healthy life. In cases in which the shunt is outside the liver (*i.e.*, congenital extrahepatic shunts), the surgeon makes an abdominal incision, finds the abnormal blood vessel and closes it off to redirect the blood flow back through the liver. To close the abnormal vessel off too suddenly risks the development of fatal portal hypertension because the vessels inside the liver may not be developed enough to handle the increased flow. For that reason, surgeons closing extrahepatic shunts may use an ameroid constrictor ring which gradually closes the shunt over a 1-2 week period as the inner ring absorbs fluid and swells to gradually close the shunt. Sutures and cellophane bands can also be used.

Unfortunately, most large-breed dogs do not have shunts located on the outside of the liver. Instead, they are affected by congenital intrahepatic (*i.e.*, inside the liver) shunts which

are much more difficult to find and correct using traditional surgeries. Locating the abnormal vessel using traditional surgery requires that the surgeon cut into the abdomen and the liver, increasing recovery time, the probability that the dog will bleed to death or that he will suffer serious damage to the liver. Mortality rates for correcting intrahepatic shunts with traditional surgery can be as high as 66%.

Interventional Radiological (IR) Surgery

Chick Weisse, VMD, DACVS, former faculty at the University of Pennsylvania's Ryan Veterinary Hospital and now at Animal Medical Center in NY, pioneered an experimental, minimally-invasive, interventional radiological (IR) surgery to correct intrahepatic shunts. Peri-operative mortality rates for this surgery are below 12%. Recovery time is minimal because there is no abdominal incision. Assoc. Professor Matthew Beal, DVM, at Michigan State University's College of Veterinary Medicine and Brian Scansen, DVM, at Ohio State University are also doing the procedure with similar outcomes. (Surgeries are also being done on the West Coast. Contact Dr. Chick Weisse at (212) 329-8816 for information.)

The IR surgical technique involves threading a catheter through the jugular vein and placing a mesh sleeve (stent) inside the vena cava where the shunt enters, having bypassed the liver. The surgeon then places several embolization coils on the shunt side of the mesh stent. The coils are coated with a thrombogenic material so that clots develop around the coils, gradually closing off the abnormal blood flow through the shunt. There is still risk of portal hypertension. As such IR surgeons err on the side of placing too few coils rather than too many. IR surgery takes as little time as 70 minutes. Moments after the dog awakes, it plays as if nothing happened. Post surgery, the dog is gradually weaned off previous medications treating the side effects of the shunt (e.g., metronidazole and lactulose) and can begin to eat normal dog food. Recovery time is minimal. Some post-surgical gastro-intestinal problems have been noted.

Implications for Puppy Buyers & Breeders **Does the litter contain a shunt puppy?**

If the answer is "yes", then BOTH parents are carriers of the fatal shunt gene and every puppy in that litter has on average a 75% chance of having either a shunt or being a carrier.

Has the stud dog (or bitch) EVER produced a shunt puppy?

If the answer is "yes", then the stud dog is a carrier of the fatal gene and if bred with your (non-carrier) bitch on average 50% of your puppies will be carriers, too. If your bitch is also a carrier, then your puppies are at even higher risk - on average 25% of your puppies will be shunt puppies and, on average, another 50% will be carriers.

Puppy buyers are advised to ask breeders these same questions when inquiring about a puppy.

Support IHPSS Research with DNA!

**We can beat this by sharing
information honestly & working together.**

How Can We Prevent IHPSS?

We do not yet have a genetic test to identify BMDs that carry the gene for IHPSS. As such, a breeder cannot tell if they own a carrier until the dog produces an affected puppy. By that time, the dog has also produced many other puppies that are carriers. The mode of inheritance in BMDs is believed by Dutch researchers to be autosomal recessive. Currently, the best way to treat liver shunts is to avoid breeding dogs that could pass on the disease trait. Affected dogs and their off-spring should not be bred. Siblings of affected dogs should only be used after careful screening. If any affected offspring are born, continued breeding of the parents should be discontinued. These cases should be recorded in the Berner Garde Database. (www.bernergarde.org)

In the meantime, breeders can screen their litters with a relatively inexpensive blood ammonia test a few weeks after birth to ascertain the possibility of a shunt. Breeders should carefully monitor failing or failed puppies.

Research Isolating the IHPSS Gene in BMDs

Dr. Jan Rothuizen, a veterinary geneticist at Utrecht University in the Netherlands, in collaboration with Dr. Robert Washabau at the University of Minnesota School of Veterinary Medicine, is attempting to find a genetic marker for IHPSS in BMDs. Isolating the IHPSS gene is the first step on our path to developing a genetic test for IHPSS carriers, similar to what we have for vWD. (NB: We have tests to identify affected dogs; we do not have tests to identify (normal looking) carriers. This study is a foundation for such a test). If you own an IHPSS-affected BMD or, as a breeder, your dam and/or sire have produced an IHPSS-affected BMD puppy, please participate in this study by contacting Dr. Rothuizen at j.rothuizen@vet.uu.nl or Nancy Melone, Ph.D., at nancymelone@mac.com. **Study participation is completely confidential.** Ideally, we would like DNA from the parents, the affected puppy and as many siblings as possible, but we will accept whatever DNA you can donate.

To participate, we will need copies of: (1) the affected dog's pedigree, (2) the diagnosis (e.g., blood test for ammonia, radiograph, surgery) and (3) 4-8 mL blood draw (EDTA-coated, purple top tube) from the affected dog and as many relatives as possible (i.e., parents, siblings). In North America, blood, pedigree and diagnosis should be sent by Express Mail to Dr Robert Washabau, Dept. of Veterinary Clinical Sciences, College of Veterinary Medicine, 1352 Boyd Ave., University of Minnesota, St. Paul, MN 55108. Notify him that a sample is coming at (612) 625-5273 or washabau@umn.edu.

About the Author - Nancy, who holds an MBA-Ph.D. in computer information technology, is a fancier of Bernese Mountain Dogs as a new breeder, competitor and contributor to breed publications. She serves on the BMDCA Breeder Education and Herding Committees and is active in canine health and research. A student in veterinary technology at Purdue University, she is an active member of Dog Writers Association of America, AKC, Canadian KC, Bernese Mountain Dog Club of America, Westmoreland County Obedience Training Club and Three Rivers Bernese Mountain Dog Club. Nancy can be reached at nancymelone@mac.com. If you have ideas for other BMD Owner Guides, please let Nancy know.