Portosystemic Shunt (Liver Shunt) in the Scottish Terrier

By Linda L. Orsborn

The Scottish Terrier is one of over 74 breeds of purebred dogs[1] reported to produce the genetic defect of congenital portosystemic shunt. Currently our STCA Health Survey reports a carrier frequency of 4.7%.[2] This is a **rare** condition and not a widespread problem in the Scottish Terrier at this time. Unfortunately genetic disorders in dogs can spread rapidly. Two factors, in particular, influence the spread of a genetic disease: 1.) a lack of understanding about the disease and its symptoms and 2.) the widespread use of carrier dogs or bitches, well respected in the area of conformation, for breeding purposes. Much is still unknown about portosystemic shunt and its mode of inheritance. Because PSS is thought to be a polygenic disease, the parents may not contribute equally to the traits responsible for affected puppies and the carrier offspring may only inherit a piece of the carrier parent's traits. A percentage of puppies produced by carrier parents will also be carriers. The purpose of this article is to deal with factor 1.) – the disease, portosystemic shunt, recognition of its symptoms, methods of arriving at a diagnosis and options for treatment.

What is a Portosystemic shunt?

Portosystemic shunts (PSS) are the result of abnormal vascular connections between the portal vein and the systemic circulation. The portal vein should connect the gastrointestinal tract with the liver. Figure 1. is an illustration of correct anatomy.

Correct or normal placement of the portal vein allows blood from the intestinal tract to flow directly to the liver. The liver then performs its function of metabolism and detoxification. The cleansed blood then returns to the heart and is pumped to the rest of the body.

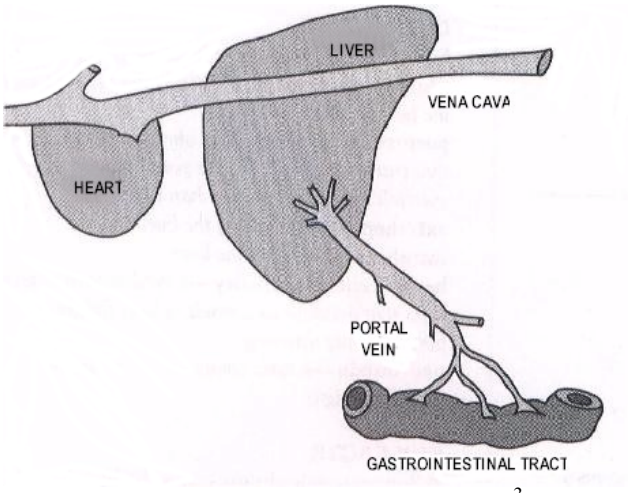

Figure 1. Correct Portal Vein Anatomy[3]

Shunts can be present at birth or acquired later in life as a result of disease process. Approximately 75% of shunts are present at birth.[4] They are the result of anatomical anomalies or failure of fetal vessels to close after birth. Shunts are classified by their location: the extrahepatic shunt is outside the liver and the intrahepatic shunt is inside the liver.

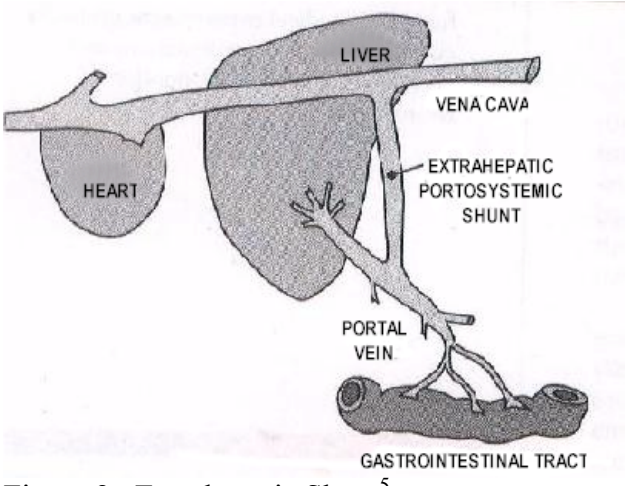

Figure 2. Extrahepatic Shunt[5]

Typically, smaller dogs, such as the Yorkshire Terrier, Maltese, Cairn Terrier and Scottish Terrier have extrahepatic shunts and larger dogs

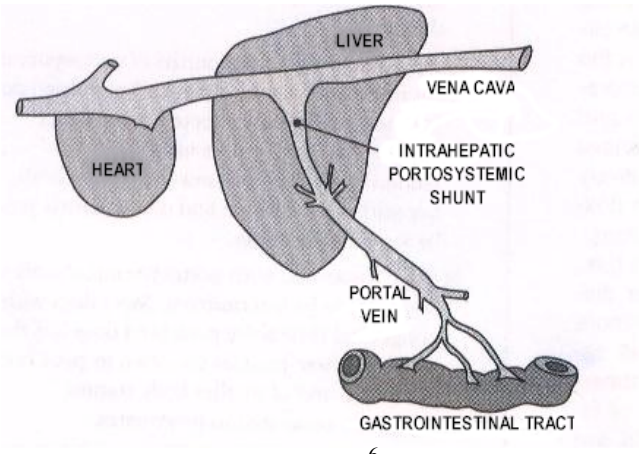

Figure 3. Intrahepatic Shunt[6]

such as Golden Retrievers and Irish Wolfhounds have intrahepatic shunts. Size exceptions have been noted within the intrahepatic classification of shunt. Extrahepatic shunts occur more frequently than intrahepatic shunts. Additionally, another phenomenon called microvascular dysplasia (MVD), a series of small intrahepatic shunts appear to be highly prevalent among Cairn Terriers. Hepatic microvascular dysplasia is thought to be a related disease to PSS that the Cairn Terrier carries. Research scientists are not of one mind about this phenomenon. A Cairn Terrier research project was undertaken by Thomas Schermerhorn, Sharon A. Center, Nathan L. Dykes, Peter H. Rowland, A. E. Yeager, H. N. Erb, Karen Oberhansley and Michael Bonda. They published an article in the Journal of Veterinary Internal Medicine, Vol. 10, No. 4 (July-August) 1996: pp 219-230. The article's title is "Characterization of Hepatoportal Microvascular Dysplasia in a Kindred of Cairn Terriers." This article is more technical than practical for our particular discussion. Within the article is a description of anatomical changes in livers of PSS Cairns and description of anatomical liver changes of MVD Cairns. MVD Cairns can appear to be healthy and they can go undiagnosed throughout their lives. If they are undetected, they often remain in the breeding population. The role of MVD Cairns in the spread of PSS isn't known. At this time we do not know if the MVD phenomenon occurs in our Scottish Terrier population.

Acquired Shunts

The differentiation of congenital and acquired shunts is important to this discussion. Acquired shunts are not hereditary in nature. They are a result of progressive liver disease. Dogs suffering from cirrhosis, hepatitis or congestive heart failure can have increased pressure inside their livers. This pressure causes embryonic vessels that normally have no function after birth to open. Often this involves a number of vessels not the one or two seen in congenital shunts. The presence of shunts in an autopsy of an older dog does not mean this dog carries the genetic traits for portosystemic shunt. It requires further examination of liver tissue. Careful examination of the dog's medical history and a study of liver tissue should reveal the origin of this dog's shunt.

Symptoms of PSS in the Scottish Terrier

There are multiple symptoms associated with this disease and the number and severity of symptoms depends on how much blood bypasses the liver. Symptoms are most often seen at a young age. The typical PSS dog is purebred and under one year of age.[7] Dogs as old as eight and ten years have been discovered to have PSS. Although recorded, this is not the norm. Affected dogs can be small in stature, unthrifty and sometimes anorexic. Coat and skin condition are poor. They develop urinary tract infections at an unusually young age. The cause of the urinary tract infection is ammonium urate crystals. These crystals are formed by excessive ammonia and uric acid in the urine.[8] PSS puppies do not react well to sedatives or anesthesia. The liver cannot metabolize these agents to help eliminate them from the dog's body. The sedatives are shunted back to the heart and returned to the body's circulation. PSS dogs can be subject to bouts of lethargy and depression. Dogs with severe shunts develop additional central nervous system signs like hyperactivity, circling behavior, disorientation, severe aggression, weakness, excessive salivation, staggering, temporary blindness, seizures and even coma. Vomiting and diarrhea are present in 2/3 of the cases of PSS. All of the PSS symptoms are affected by the

consumption and digestion of protein rich foods. These foods produce ammonia, gamma-aminobutyric acid, natural benzodiazepines and mercaptains during the process of digestion. These chemicals are potential neurotoxins.[9]

Breeder Observations

As the author of this piece my intention is to present information for other breeders and owners to use if necessary. Opinion and emotion can obstruct this purpose, but observation of fellow breeders can be helpful. During the course of research on this disease I spoke to Yorkshire Terrier breeders, Cairn breeders and Scottie breeders. One scenario was repeated to me several times. It mirrored my personal experience. This narrative is anecdotal and not intended to imply that all cases follow this pattern.

A litter of puppies is born and progresses normally. By six weeks of age the puppies are completely weaned and all appears to be well. During the next four to eight weeks the puppies increase their consumption of protein rich puppy food. At this stage of life the introduction of vaccines becomes the next step in the successful raising of the litter. Between six weeks and fourteen weeks most puppies are given three to four sets of vaccinations. Some are combination vaccines and others are individual vaccines. (As a breeder I have never experienced puppies with a vaccine reaction but, I have read the articles concerning the possibility of adverse vaccine reaction especially with combination shots.)

One night my litter was healthy. The next day one puppy, Holly, was wobbly, disoriented and clearly in distress. The outside pen was checked for leaves or other foreign material the puppy could have ingested. Electric cords were checked in the den. All the cords had been raised out of the puppies' reach. We checked Holly for bites or a possible bee sting. Truthfully, it was too early in the season for bees in Massachusetts. Next her temperature was taken. It was normal. Nothing external appeared to have happened to this nine-week-old puppy. Why was she suddenly so sick? Off we went to the veterinarian.

Dr. Cindy Shaefer had seen Holly and her sisters for a routine vaccination and checkup the week before. Dr. Shaefer ordered blood work on Holly and examined her. Nothing specific was found. Later in the day Holly was almost normal in behavior. Then later at night her behavior was nothing less than bizarre and she staggered like a drunken sailor. By morning she was again nearly normal. The blood chemistry results were all within normal range and the blood count showed no sign of infection. For two days this erratic pattern of close to normal behavior and periodic staggering accompanied by rages or vacant staring persisted. Then Holly had a mild seizure. We consulted a veterinary neurologist. He found her symptoms, at the time of examination, unremarkable. She became worse and the seizures and behavior changes occurred more frequently. At the second visit to the neurologist he stated two possible causes for Holly's illness vaccine reaction or possible brain malformation. Some other breeders have reported the initial concern by veterinarians about vaccines.

Neurological tests like MRI and a spinal tap were discussed as possible options for determining the cause of Holly's illness. The first logical course of action was a bile acid test, a simple inexpensive blood test. The neurologist did not strongly believe that Holly was a PSS puppy; however, the test would rule out shunt and tell us if Holly could tolerate Phenobarbital for her seizures. The bile acid test detects diminished liver function. Dogs on seizure medication and older dogs with liver failure symptoms often require bile acid tests to monitor their health status.

For the young dog high bile acid results indicate a need for a PSS work up. This test is an effective **screening test**. It does not provide definitive diagnosis. Rather, it tells you if more testing in this area is needed. Breeders in other breeds report spending thousands of dollars on sophisticated neurological tests only to find that the simple fifty dollar test, the bile acid test, provides them with a direction for diagnosis of their sick puppies.

Diagnosis of a PSS Scottish Terrier

Screening Test – The bile acid test – Bile acids are produced by the liver from cholesterol. This is work. In general, animal bodies hate to do extra work. So bile acids are recycled in the body. They do their job and are released into the intestinal tract. They are reabsorbed by the digestive tract into the circulation. The liver recycles them rapidly. Therefore, there shouldn't be high levels in the blood stream at any time and there isn't much difference in levels before and after meals. High levels of bile acids indicate that the liver can't handle the recycling job due to damage or diminished capacity.[10] The bile acid test is a simple one. A blood sample is drawn after a twelve hour fast. The dog then receives a small meal of high protein food. Two hours later a second blood sample is drawn. These samples are analyzed for bile acid content.[11] The fasting range should be 0.0 – 5.0 umol/L and the two-hour sample should be 5.0 – 25.0 umol/L. PSS puppies have marked high numbers. As an example, Holly's numbers were 179.2 umol/L fasting and 219.9 umol/L two hours after food. If you have a puppy with high bile acids, it is time to consider the possibility of more sophisticated tests.

Diagnostic Tests – The GDC (Institute for Genetic Disease Control in Animals) accepts the following four procedures for definitive diagnosis of PSS:

- Ultrasound
- Scintigraphy
- Surgery
- Necropsy

Ultrasound – As little as ten years ago this technique was not widely used. Today it has emerged as one of the most popular imaging modalities. It is safe, requires little or no sedation, is readily available and offers much information. However, it is time consuming and is dependent on the skill of the sonographer. Ultrasound often shows small liver size consistent with a shunt. Occasionally, large kidneys and urinary tract calculi may also be identified to support the presence of a shunt. Doppler ultrasound studies of the vessels may be used to detect abnormal blood flow. Doppler ultrasound can sometimes detect turbulence in the caudal vena cava, which is circumstantial evidence of a shunt.[12]

Transcolonic Portal Scintigraphy – Nuclear medicine is an effective way to detect a portovascular anomaly. Scintigraphy requires minimal or no sedation and the patient is required to remain still for only two minutes. The need for special equipment and handling of radioactive material limits the availability of this test.[13]

To image the portal system, a small concentrated volume of techetium pertechnetate is infused into the colon. This radioisotope rapidly absorbs across the colon wall into the left colonic vein that drains into the portal vein. In the normal dog the isotope goes from the colon to the liver to the heart. In PSS dogs the flow is colon to heart to liver. Sometimes the flow arrives simultaneously in the heart and liver.[14] This test confirms a shunt, but not necessarily its location. The advantage of scintigraphy is its non-invasive nature of determining the presence of a shunt. Its disadvantage is the lack of availability. The dog owner may have to travel great distances to locate a center with the proper equipment.

Surgery – Surgery for PSS is not undertaken at your local veterinary clinic. It is a delicate and specialized procedure done at hospitals connected to veterinary colleges or large research centers. If your puppy has high bile acids and other consistent symptoms, you can elect to pursue surgery as soon as possible. A workup will be done to determine if the puppy is a good candidate for surgery and is in condition to withstand the surgery.

A mesenteric portogram is the contrast study of choice at this time. A laparotomy is performed to exteriorize a loop of jejunum and omentum. A catheter is placed in a jejunal vein. The abdomen is temporarily closed. Contrast media is hand injected through tubing connected to the catheter and an exposure made at the end of each injection. Several different views are taken. The liver is biopsied and portal pressures are measured from

the catheter.[15] If tests confirm a repairable shunt the corrective surgery is usually done at this point because the dog is already under general anesthesia.

In the past, shunts were ligated. Portal pressure was monitored and the shunts were closed down as much as possible. It was often not feasible to completely close the shunt. Too much blood flow returned to a small or damaged liver could result in sudden death. It was hoped to achieve at least a 60 to 80% reduction of improper blood flow. Dogs with lesser percentages of ligation had ongoing symptoms and their long-term prognosis was guarded.

Today, the preferred surgical technique for single extrahepatic shunts is the use of an ameroid constrictor. Single extrahepatic shunts closed with the ameroid have a 75% success rate.[16] The ameroid is a ring lined with an absorbent material. Over a four to six week period the material inside the ring expands and slowly shuts off the blood flow in the shunt.

Recovery from the surgery has a range of success. Much is dependent upon the damage the dog may have sustained before surgery. Some dogs have experienced severe seizure activity during the course of illness and the seizures and neurological symptoms can persist after surgery. On the other hand, if the percentage of normal blood flow restored to the liver is high, the dog may be healthy. A lifetime diet of low protein food like Hills k/d will be necessary for the repaired PSS dog. These dogs do have an increased risk of developing bladder stones, but the quality of life can be excellent for some repaired PSS dogs.

It is not responsible to discuss the option of surgery without at least mentioning cost. This is not an inexpensive disease to treat. Estimates well in excess of two thousand dollars can be expected for diagnosis and surgery. In Holly's case, post surgery complications nearly doubled that figure. For some breeders and pet owners the cost of surgery is simply not affordable.

Necropsy - Portosystemic shunt in the Scottish Terrier is believed to be a polygenetic trait. Research points to its genetic basis, but has not discovered the mode of inheritance or the genes involved. If you think a dog in your kennel may be a PSS dog, you need to confirm or exclude the diagnosis. If the dog expires and you have not done confirming tests, have the dog's liver examined for changes consistent with shunt. This may require that you send samples away to a veterinary college or research facility instead of having your own veterinarian conduct the autopsy. Not knowing or guessing that you have a deadly disease in your breeding stock is foolish. Have your puppies diagnosed, even the dead ones. It is the only logical course of action. If you have a PSS puppy, please consider registering him/her with the GDC.

Medical Treatment for PSS

We have covered symptoms and diagnosis. Medical treatment, by itself, is not a long-term treatment of choice. One should not expect to keep a dog on medical treatment without the prospect of surgery. Quality of life would be poor. However, antibiotic treatment with neomycin does help lessen the neurological symptoms. Neomycin is an antibiotic that is not absorbed by the digestive tract. It will kill the bacteria in the intestines that produce ammonia. This reduces the neurotoxins in the body. It is often used in conjunction with Lactulose. Lactulose converts ammonia to ammonia ions. The ions are not readily reabsorbed into the body. These drugs help to suppress the affects of ammonia on the nervous system. To further reduce the problem of ammonia, a low protein diet is fed to these dogs. Dogs with inoperable shunts and dogs waiting for surgery follow this protocol. After surgery the hope is that a low protein diet alone will be sufficient to maintain the dog in good health. A common food used for this purpose is Hills k/d.

How significant is Portosystemic shunt? Should Scottish Terrier breeders and owners be concerned?

The guesstimate rate of carrier frequency of 4.7% for PSS is a small number of Scottish Terriers when compared to the frequency for Scottie Cramp, skin diseases or hypothyroidism. This Bagpiper will be delivered to over seven hundred homes. I believe that the number of readers who will recognize PSS as a problem in your own breeding program will probably be well under two dozen. (I have found only nine identifiable PSS puppies in the last eight years.) Perspective is important. This disease is rare in our breed, but it is considered by many to be epidemic within the Cairn and Yorkshire Terrier populations. Knowledge is power. We need to educate ourselves about genetic disease and always diagnose our sick puppies. Disease can spread if we are not informed and aware of the genetic potential in our own kennels.

CMO is also a rare disease. Its guesstimate of carrier frequency rate in the Scottish Terrier is 4.7%,[17] the same frequency rate as portosystemic shunt in the Scottish Terrier. The mode of inheritance may be different and actual numbers of affected puppies may be different, but the principle is the same. Our Scottish Terrier breeders, the STCA Board of Directors, the STCA Health Trust Fund Trustees, members of the STCA Health Education Committee became aware of the suffering that CMO can cause. They learned that it can be spread into our Scottie population and acted to fund a research project in conjunction with the Cairn Terrier Club of America and the West Highland White Terrier Club of America. Hopefully a DNA test for detecting CMO carriers will be developed within the next two years. We've already accomplished the accepted use of a DNA test for the detection of carrier, clear and affected status for vWD. Now we need to at least educate ourselves about PSS. It is a painful, potentially deadly and costly disease – a very unfortunate way for any Scottish Terrier to live or die.

What should we do today?

If you produce a PSS puppy, accept the fact that **both** the sire and the dam are now proven carriers. Start pedigree tracking from this day forward. Other puppies produced by the dog or bitch could be carriers. Currently, there is no way to test for carriers. Keep accurate records. DNA samples of the affected puppy, its littermates and parents can be taken and stored. The Cairn and the Yorkshire Terriers are already working on a possible DNA project with scientists from Michigan State University and Ohio State University. Register your PSS puppy in the GDC Terrier PSS research registry. The results are not open to the public, but will be used for research purposes. Today, Eowyn's Holly Go Lightly is the only Scottish Terrier in this registry. Hopefully, this article will prompt the registration of more affected Scottish Terriers.

Bile acid test the littermates of an affected puppy to ensure they have normal liver function. We must not sell affected puppies to unsuspecting pet buyers nor do we want them in our breeding programs. Remember that a percentage of affected dogs show no symptoms of PSS until they are well into adulthood. Be informed and share your knowledge. Scientific advances against genetic diseases are happening every day.

To reach the GDC to inquire about the PSS research database call (503) 756-6773 or visit them at http://www.vetmet.udavis.edu/gdc.html

Linda L. Orsborn

End Notes

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