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Portosystemic shunt – persistent ductus venosus – in the Irish Wolfhound

INTRODUCTION

Circulation leaving the stomach, intestine, spleen, and pancreas enters the portal vein and flows through the liver before returning to the systemic circulation. Blood leaving the gastrointestinal tract is containing nutrients, hormones, toxins and bacteria, which normally are removed by the liver. When some of these substances escape into the systemic circulation, as in the case of portosystemic shunts (PSS), clinical signs of hepatic disease appear.
The relationship of portal blood flow to normal hepatic function is also very important.

Portosystemic shunts (PSS) are abnormal vessels that directly communicate between the portal vein and vessels in the systemic circulation. When portal blood bypasses the liver, potential toxins reach the systemic circulation, and the liver also is deprived of hepatotrophic substances. This results in various degrees of hepatic encephalopathy (nervous symptoms), atrophy of the liver and/or other signs of hepatic insufficiency.

PSS is a relative common phenomenon in companion animals, and may be congenital or acquired. Congenital PSS may be classified as intrahepatic or extrahepatic shunts on the basis of location. Cats and smaller breed dogs usually develop single extrahepatic shunts, whereas single intrahepatic shunts are more commonly seen in large breed dogs. Multiple extrahepatic shunts are believed to be acquired secondary to pre-existing liver disease.

Congenital PSS may occur in any breed. However, clinical reports suggest increased incidence in certain breeds. No sex predisposition is apparent. Most animals develop signs before one year of age, but congenital PSS have been recognised in dogs ten years of age.

In the Irish Wolfhound a persistent ductus venosus is diagnosed. The ductus venosus is a fetal vessel providing a shunt for blood from the gastrointestinal tract and into the umbilical vein to bypass the liver before birth. This connection is normally closed within 60 hours after birth in the dog, causing the entire portal blood flow to perfuse the liver. Failure of the shunt to close in some animals is suggested to result from several abnormalities.

CLINICAL SIGNS

Clinical signs are highly variable, but referable to the central nervous system, the gastrointestinal system and/or the urinary tract. The most common clinical signs are associated with hepatic encephalopathy, such as ataxia, depression or stupor, unusual behaviour or disorientation, and seizures. These signs often wax and wane and may be exacerbated when food containing high levels of protein is eaten or by gastrointestinal bleeding. This because ammonia, one of the most important toxins implicated, is derived from bacterial deamination of amino acids in the intestine. The gastrointestinal signs usually are mild and intermittent, including anorexia, vomiting/diarrhoea, pica and sometimes polyphagia. Other sighs include delayed anaesthetic recovery, polydipsia/polyuria, dysuria and hematuria resulting from urate calculi.

Physical examination findings usually are non-specific, but most individuals show stunted growth and failure to gain weight.

DIAGNOSIS

Tentative diagnosis of PSS is based on history, clinical sighs and results of laboratory evaluation. Several hematological and biochemical abnormalities can occur. Individual parameters may however be only mildly abnormal, but the overall results will reflect a pattern suggesting hepatocellular dysfunction. The serum bile acid (SBA) test, including fasting and 2 hours postprandial serum bile acid determination, is generally regarded as the screening test of choice in ordinary practice. A high fasting serum ammonia concentration and the ammonia tolerance test are also useful biochemical tests in identification of PSS.

The bile acids are produced in the liver and secreted into the biliary system during feeding. In the intestine they take part in the digestion of food. They are normally absorbed primarily in the lower part of the intestine, the ileum, where transportation back to the liver takes place in the portal vein.

The fasting concentration of SBA may be normal or only mildly elevated. It will increase shortly after feeding, but is normalised within two hours in healthy individuals. In patients with PSS, where portal blood is bypassing the liver, the concentration will be much higher than normal.

Urinary sediments may be examined for ammonium biurate crystals. These crystals are often observed, but are not pathognomonic for a shunt, nor dooes an absence rule out the possibility of a shunt.

Plain abdominal radiographs may reveal a small liver, large kidneys and sometimes urinary calculi. The shunts are identified radio-graphically by angiography, a positive contrast radiography, or by ultrasonography.

TREATMENT

A symptomatic treatment, consisting of a diet low in protein and medical management may be performed. In larger breeds the result generally is not satisfying.
The only real therapy is surgical ligation of the portosystemic shunt vessel. This is a complicated procedure, especially when the shunt is intrahepatic, as in the Irish Wolfhound.

THE NORWEGIAN SHUNT PROJECT

Because of an apparently high prevalence of PSS in the Irish Wolfhound compared to other breeds in Norway, the Norwegian Irish Wolfhound Society in co-operation with the Department of Small Animal Clinical Sciences, in 1988 decided to examine all Wolfhound puppies at the age of seven to eight weeks. The breed society now requires screening of the puppies before recommending them for sale.

The screening consist of the SBA-test and biochemical examination of other serum liver parameters of current interest. In some puppies urinary sediment is examined for ammonium biurate crystals. The blood and urine samples are mailed to the Veterinary College, where they are analysed. The samples have to be accompanied by the dogs pedigree. When PSS is suspected the diagnosis is confirmed by splenoportography.

The breeder gets a certificate which include the dogs identification, number of dogs tested (litter size) and number of dogs with possible PSS. This certificate is delivered to the new owners when the puppies are sold.

So far 54 Norw4egian litters are tested and PSS has been identified in 12 of them. One or two individuals in each litter have been affected. Some of the puppies have been “poor doers”, with poor appetite, smaller and not as active as other dogs in the litter, while others have not shown any symptoms at the age of testing.

Since 1984 about 300 Norwegian Irish Wolfhounds from seven weeks up to two years of age, have been tested. PSS has been diagnosed in 18 dogs. According to results of the screening test, another three dogs have been suspected, but confirming the diagnosis has not been made possible. The anomaly has also been diagnosed in at least five Swedish Wolfhounds tested in Norway. Some of the affected dogs are related, and it seems to be an overrepresentation of PSS in certain lines. Because of a probable breed and familial disposition for this anomaly have given offspring with PSS and to control breeding, the mode of inheritance is of interest. Test matings and further evaluation of pedigrees will be carried out, and hopefully will clarify the question of inheritance. So far the breeders are advised to avoid breeding dogs that of littermates.