PORTOSYSTEMIC SHUNTS (PSS) IN THE IRISH WOLFHOUND

Introduction

The liver is one of the largest organs of the body, situated in the abdomen directly behind the diaphragm. The liver plays a crucial role in the metabolism of proteins, fats, and carbohydrates, but also in the detoxification of waste products and xenobiotic compounds. The liver receives most of its blood from the portal vein, via which a lot of this waste from the intestines reaches the liver for detoxification. The major waste product is the very toxic ammonia, which is a by-product of the intestinal protein breakdown.

Portosystemic shunts

A portosystemic shunt (PSS, also called liver shunt) is a vessel, which is normally not present after birth and which runs between the portal vein and the systemic venous circulation. Thus the intestinal blood does not enter the liver but bypasses it and reaches the systemic circulation without hepatic detoxification. High levels of ammonia and other toxins enter the body system and a plethora of symptoms may occur, among which gastrointestinal signs, urological problems, weight loss, and signs of cerebral dysfunction (e.g., apathy, circling, apparent blindness, seizures, and coma) are frequently seen.

Multiple portosystemic shunts may develop secondary to chronic liver disease, but mostly a PSS is a single vessel. In other words, a PSS is an extra vessel which is there from birth on, and is thus to be seen as a congenital developmental disorder. Grossly, two types of congenital PSS are seen (Fig. 1): the ones within the liver (so-called intrahepatic shunts=IPSS), and the ones outside the liver (extrahepatic shunts=EPSS). As a rule of thumb, IPSS are seen in larger dog breeds, such as the Irish wolfhound, retrievers, Bernese mountaindog, etc. and EPSS are frequently seen in small dog breeds, such as the Dachshund, Yorkshire terrier, Cairn terrier, etc.. The observation that certain types of PSSs prevail in certain breeds was the first indication that congenital PSS is a hereditary disorder. Since 1983, several of Netherlands' breeders' associations (i.e., the breeders's associations of the Irish wolfhound, Cairn terrier, and deerhound) of affected breeds have started a screening program for PSS.

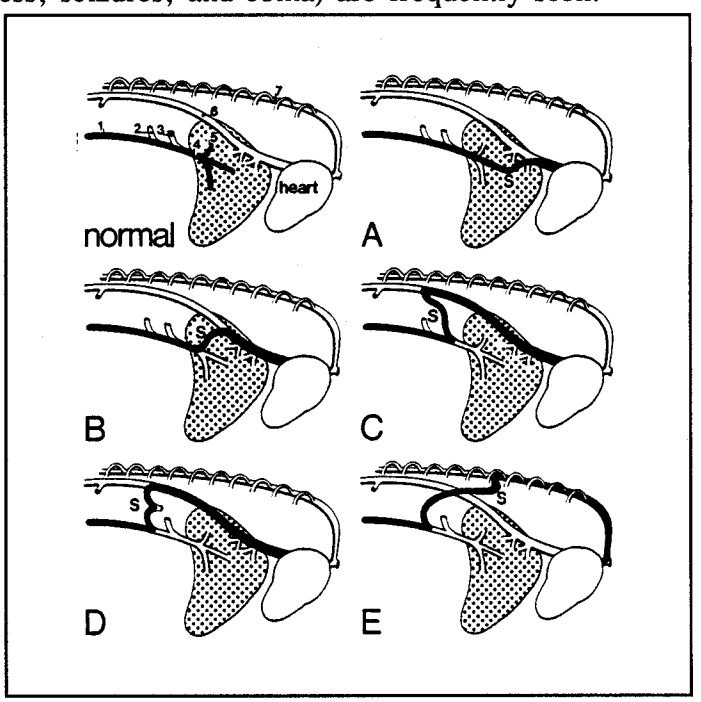

Figure 1. Diagram showing the normal portal circulation and various types of intrahepatic (A-B) and extrahepatic (C-E) portosystemic shunts.
1 = portal vein
6 = main systemic vein

Screening for PSS

The purpose of a screening program for PSS is twofold. First, data on the prevalence of the disorder and pedigree analysis will help to unfold the genetic basis of the disease and possibly to set up a breeding program to eradicate the disorder. Second, by screening every litter a future owner will get a guarantee that his pet does not suffer from a liver shunt. The screening for PSS relies on the circulation of increased quantities of toxins and by-products in the systemic blood stream. Of these, the fasting venous (or arterial) ammonia concentration is the most reliable. Disadvantage of the ammonia determination is that the incidence of falsely high results may be high due to mistakes during sampling procedure (e.g., contamination of the sample with saliva, sweat, or smokers' breath and insufficient cooling or prolonged storage). Also, the ammonia measurements throughout one breed should be done by the same laboratory procedure (so-called enzymatic method; reference values for the fasting venous ammonia concentration (Utrecht): 22-46 μmol/l). Otherwise the results of the litters cannot be compared.

Hence the ideal situation is to perform all ammonia determinations in one specialized veterinary center. Since Holland is quite small, all Irish wolfhound litters have been tested at the University Clinic for Companion Animals in Utrecht since 1983. In larger countries, this seems not feasible, so in these situations another screening method seems warranted. Unfortunately, all other measurements are less sensitive in detecting PSS than determination of the ammonia concentration, and are thus prone to false negative results. Of the alternatives, determination of the total bile acids in plasma is the best, because sample handling is less critical than for ammonia. However, 38% of the dogs with a PSS have normal fasting bile acids concentrations, so a considerable per cent of affected dogs will be missed by screening bile acids only. Also, in case of high bile acid concentrations, the diagnosis PSS has always to be confirmed by other means, since high bile acid concentrations are not specific to PSS.

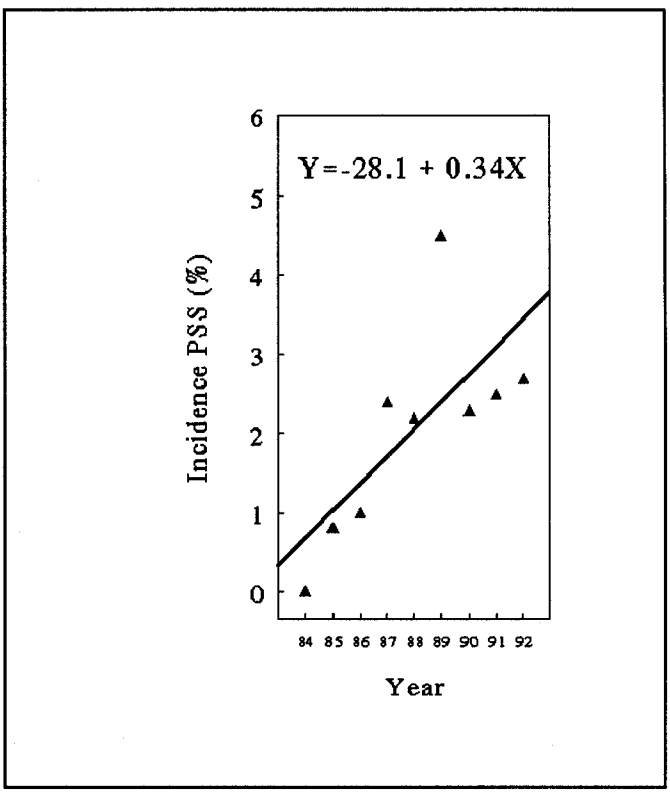

Figure 2 Incidence rate of PSS in Irish wolfhound plotted per year

Results of the screening in Irish wolfhounds

Since 1983, 1066 Irish wolfhound pups have been tested at the age of 7-8 weeks. The average incidence rate of intrahepatic PSS in that period was 2.0%, which is significantly higher than in the general dog population. The incidence rate increased with 0.34% per year (Fig. 2). These data underscore the genetic basis of PSS in the Irish

wolfhound. This is further proven by the difference in coefficients of relationship between the dogs with PSS and the healthy control Irish wolfhounds.

Surprisingly, the mean ammonia concentration in clinically normal Irish wolfhound pups (74 μmol/l) was higher than in the general population (33 μmol/l) (Fig. 3). Thus far this phenomenon has not been explained.

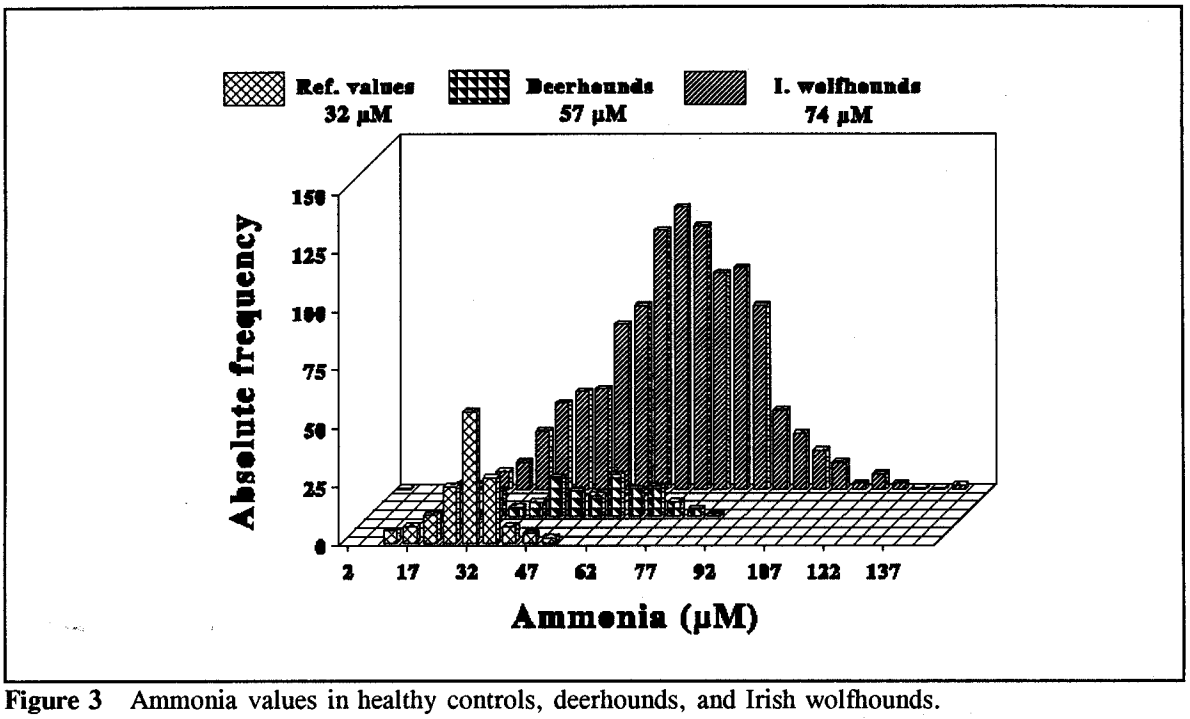

Figure 3 Ammonia values in healthy controls, deerhounds, and Irish wolfhounds.

Conclusions

Continuing the testing will ensure the owners that they have bought a dog without PSS and will eventually lead to elucidation and hopefully eradication of PSS in the Irish wolfhound. This screening program is an excellent example of the usefulness of tight cooperation between a breeders' association and the veterinary profession in order to take adequate steps against genetic disorders. Further research is needed to elucidate the questions that have risen during the presented investigations.

H.P. Meyer, DVM

Department of Clinical Sciences of Companion Animals,
Faculty of Veterinary Medicine, Utrecht University
PO Box 80.154
NL-3508 TD Utrecht
the Netherlands
fax 31-30-518126