

# Association of breed with the diagnosis of congenital portosystemic shunts in dogs: 2,400 cases (1980–2002)

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**Objective**—To determine the annual and overall proportion of diagnoses of congenital portosystemic shunts (CPSS) in dogs and identify breeds at increased risk for CPSS.

**Design**—Retrospective study.

**Animals**—2,400 dogs with CPSS from veterinary teaching hospitals that reported to the Veterinary Medical Database (VMDB) from January 1, 1980 to February 28, 2002.

**Procedure**—The proportion of diagnoses of CPSS was calculated for all dogs and each breed recorded in the VMDB annually and for the 22.2-year period. Odds ratios and adjusted confidence intervals were calculated for breeds with at least 100 accessions by comparing odds of each breed with a diagnosis of CPSS with that of mixed-breed dogs.

**Results**—Congenital portosystemic shunts were reported in 0.18% of all dogs and 0.05% of mixed-breed dogs. The proportion of diagnoses of CPSS increased from 5 in 10,000 dogs in 1980 to 5 in 1,000 dogs in 2001. Yorkshire Terriers had the greatest total number of diagnoses of CPSS. Thirty-three breeds were significantly more likely to have a diagnosis of CPSS, compared with mixed-breed dogs. The greatest proportions of diagnoses were found in Havanese (3.2%), Yorkshire Terriers (2.9%), Maltese (1.6%), Dandie Dinmont Terriers (1.6%), and Pugs (1.3%).

**Conclusions and Clinical Relevance**—Certain breeds appear to be at increased risk for CPSS, compared with mixed-breed dogs. The increased odds ratios among specific breeds support the hypothesis of a genetic predisposition for CPSS. Clients and veterinarians should consider appropriate diagnostic tests for dogs with clinical signs and those used for breeding from breeds with increased risk of CPSS. (*J Am Vet Med Assoc* 2003;223:1636–1639)

Despite increasing information regarding the pathophysiology, diagnosis, and treatment of congenital portosystemic shunts (CPSS), little is known about the cause of this disease. Development of congenital defects may be influenced by environmental or genetic factors alone or in combination. When the prevalence of disease is higher in an individual breed, a genetic influence is suspected.<sup>1</sup> Because the annual prevalence of the disease has increased, CPSS are considered to be hereditary in Yorkshire Terriers.<sup>2</sup> Increased incidence and familial tendencies for CPSS have also been detect-

ed in Irish Wolfhounds.<sup>3–5</sup> Because of overrepresentation of Maltese, Australian Cattle Dogs, and Golden and Labrador Retrievers with CPSS in population surveys, a genetic influence has been proposed in these breeds as well.<sup>6,7</sup> Identification of breeds at increased risk for CPSS can increase a veterinarian's index of suspicion, resulting in greater likelihood of diagnosis. Additionally, evidence in support of a genetic link helps veterinarians decide whether to counsel owners on breeding dogs with a family history of CPSS.

The purpose of the study reported here was to determine the annual and overall proportion of diagnoses of CPSS in dogs and identify breeds at increased risk for CPSS.

## Criteria for Selection of Cases

The Veterinary Medical Database (VMDB)<sup>a</sup> has compiled information on diseases diagnosed in dogs and other animal species from veterinary teaching hospitals in North America. The VMDB was searched for records of all dogs with a diagnosis of single CPSS from January 1, 1980, to February 28, 2002. Search terms included CPSS, congenital portal caval shunt, congenital defect portal vein, congenital atresia portal vein, and congenital portoazygous shunt.

## Procedures

Each accession included identification number, age, sex, breed, diagnosis, and year of diagnosis. Total number of accessions and number of accessions with CPSS were recorded only once for each individual dog per year. Annual proportion of diagnoses of CPSS, defined as the number of individual dogs with CPSS accessed during the year divided by the total number of individual dogs accessed during the same year, was calculated for all dogs. Proportion of diagnoses of CPSS for the 22.2-year period was calculated for all dogs and for each breed by combining annual data.

**Statistical analyses**—Breeds with 100 or more accessions and diagnosis of at least 1 dog of that breed with CPSS were compared with mixed-breed dogs by use of a  $\chi^2$  test.<sup>b</sup> Odds ratios (ORs) were calculated, and confidence intervals were adjusted to the level of 99.9995% for 106 individual comparisons by use of the Bonferroni method.<sup>8</sup> Odds ratios for each breed were considered significantly greater or less than the reference population if the adjusted confidence interval did not include 1.00. Exact confidence limits around the OR were reported when 1 or more expected cell counts in the  $2 \times 2$  table were  $< 5$ ; otherwise, asymptotic confidence limits were reported.

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## Results

There were 1,334,841 dogs registered in the VMDB from January 1, 1980 to February 28, 2002, and 2,400 (0.18%, or 1.8 of every 1,000 accessions) of those had CPSS. The ratio of affected females-to-males was 1.14:1. Age distribution included 0 to 2 months (1%), 2 to 6 months (31%), 6 to 12 months (22%), 1 to 2 years (18%) 2 to 4 years (16%), 4 to 7 years (8%), 7 to 10 years (2%), and  $\geq 15$  years (< 1%). Age was not reported in 1% of affected dogs.

Annual proportion of diagnoses of CPSS in the VMDB increased from 0.05% in 1980 to 0.5% in 2001 (Fig 1). Congenital portosystemic shunts were reported in 110 of 201 (55%) breeds of dog, and in 169 of 331,234 (0.05% or 5 of every 10,000 accessions) mixed-breed dogs. Breeds that contributed the greatest percentage of CPSS accessions included Yorkshire Terrier (483 [20.1%]), Miniature Schnauzer (244 [10.2%]), mixed-breed dog (169 [7.1%]), Shih Tzu (119 [5%]), Maltese (100 [4.2%]), Labrador Retriever (87 [3.6%]), Golden Retriever (86 [3.6%]), Cocker Spaniel (75 [3.1%]), Pug (75 [3.1%]), and Shetland Sheepdog (59 [2.5%]). Diagnosis of CPSS in Yorkshire Terriers increased dramatically over the study period; in 1980, 8.6% of CPSS accessions were Yorkshire Terriers, whereas in 2000, that number increased to 31.6%.

Breeds with significantly greater overall proportions of diagnoses of CPSS, compared with mixed-breed dogs were determined (Table 1). The 10 breeds with the greatest proportions of diagnoses of CPSS over the 22.2-year period included the Havanese (3.2%; OR, 64.9), Yorkshire Terrier (2.9%; OR, 58.7), Maltese (1.6%; OR, 32.0), Dandie Dinmont Terrier (1.6%; OR, 31.7), Pug (1.3%; OR, 26.2), Skye Terrier (1.2%; OR, 22.9), Miniature Schnauzer (1.0%; OR, 19.8), longhaired Chihuahua (0.9%; OR, 18.7), Scottish Deerhound (0.9%; OR, 17.6), and Standard Schnauzer (0.8%; OR, 16.1). Congenital portosystemic shunts were reported in 2 breeds with < 100 accessions, including the Hovawart (1/5 dogs) and Tennessee Treeing Brindle Coonhound (1/38).

Table 1—Proportion of diagnoses of congenital portosystemic shunts in dogs accessed from the Veterinary Medical Database from January 1, 1980, to February 28, 2002

Breed	No. of affected dogs (%)	Reference hospital population	Odds ratio	Adjusted confidence interval*
Mixed	169 (0.05%)	331,234	1.0	NA
Havanese	6 (3.2%)	187	64.9	8.9–234.3
Yorkshire Terrier	483 (2.9%)	16,538	58.7	42.9–80.2
Maltese	100 (1.6%)	6,231	32	20.2–49.8
Dandie Dinmont Terrier	4 (1.6%)	251	31.7	2.3–140.1
Pug	75 (1.3%)	5,681	26.2	15.7–42.5
Miniature Schnauzer	244 (1%)	24,411	19.8	14.0–28.0
Standard Schnauzer	36 (0.82%)	4,404	16.1	8.0–30.0
Shih Tzu	119 (0.78%)	15,274	15.4	10.1–23.4
Bernese Mountain Dog	14 (0.76%)	1,825	15.1	4.7–36.8
Bichon Frise	32 (0.67%)	4,747	13.3	6.3–25.4
Cairn Terrier	27 (0.54%)	4,990	10.7	4.7–21.2
Irish Wolfhound	13 (0.5%)	2,594	9.9	2.9–24.7
Longhaired Dachshund	8 (0.5%)	1,602	9.8	1.9–30.0
Jack Russell Terrier	13 (0.43%)	3,025	8.5	2.5–21.2
Pekingese	33 (0.36%)	9,116	7.1	3.7–13.8
Miniature Pinscher	7 (0.36%)	1,964	7.0	1.2–22.7
West Highland White Terrier	25 (0.31%)	8,163	6.0	2.6–12.2
Pomeranian	27 (0.29%)	9,411	5.6	2.7–11.6
Lhasa Apso	37 (0.28%)	13,359	5.4	2.9–10.2
Old English Sheepdog	19 (0.26%)	7,189	5.2	1.9–11.4
Shetland Sheepdog	59 (0.26%)	22,348	5.2	3.1–8.8
Chihuahua	25 (0.25%)	10,041	4.9	2.3–10.3
Scottish Terrier	14 (0.23%)	6,207	4.4	1.4–10.8
Miniature Dachshund	17 (0.2%)	8,699	3.8	1.3–8.7
Standard Poodle	22 (0.17%)	12,645	3.4	1.6–7.5
Toy Poodle	33 (0.16%)	20,833	3.1	1.6–6.0
Beagle	27 (0.16%)	17,328	3.1	1.5–6.3
Samoyed	14 (0.15%)	9,476	2.9	1.1–7.6
Golden Retriever	86 (0.14%)	63,466	2.7	1.7–4.2
Cocker Spaniel	75 (0.14%)	55,165	2.7	1.6–4.3
Doberman Pinscher	53 (0.14%)	38,428	2.7	1.5–4.6
Siberian Husky	18 (0.13%)	14,084	2.5	1.1–5.9
Labrador Retriever	87 (0.1%)	90,902	1.9	1.2–2.9

\*99.9995% Confidence interval adjusted for 106 comparisons with mixed-breed dogs.  
NA = Not available.

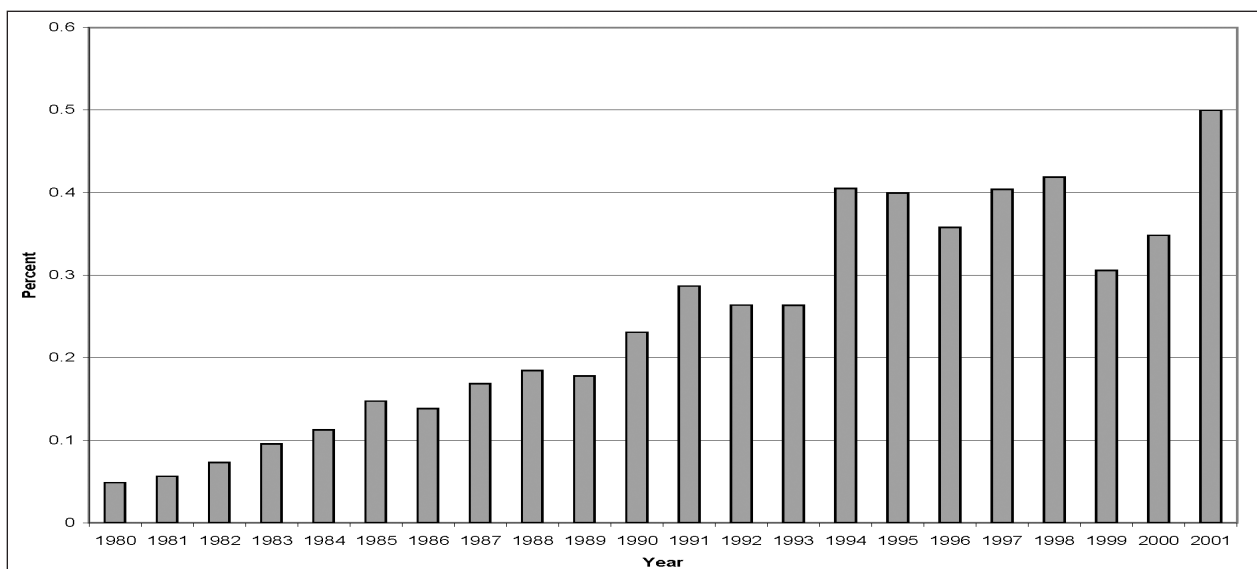


Figure 1—Proportion of diagnoses of congenital portosystemic shunts in dogs accessed from the Veterinary Medical Database from January 1, 1980, to December 31, 2001.

Odds ratios for CPSS were significantly higher for 33 breeds, compared with mixed-breed dogs. Additional breeds with ORs > 10 included the Shih Tzu (OR, 15.4), Bernese Mountain Dog (OR, 15.1), Bichon Frise (OR, 13.3), Lakeland Terrier (OR, 12.3), Cairn Terrier (OR, 10.7), and Italian Greyhound (OR, 10.5). Odds ratios for Skye Terriers, longhaired Chihuahuas, Scottish Deerhounds, Lakeland Terriers, and Italian Greyhounds were not significantly different from that of mixed-breed dogs. No breeds had significantly lower ORs, compared with the reference population.

## Discussion

Classification of a disease as inherited can be difficult when the disease is rare in the population and phenotypic appearance of the disease is not easily recognizable. A genetic contribution to disease expression is suspected when the reported frequency is higher in 1 breed than in others, environmental factors common to members of the breed are insufficient to account for the observed differences in frequency, and the same or a similar disorder is proven to be inherited in another species of animal or in humans.<sup>1</sup> Through studies of prevalence and evaluation of family groups, single CPSS, or closely related diseases such as microvascular dysplasia, have been identified as hereditary in Yorkshire Terriers, Irish Wolfhounds, Cairn Terriers, and humans and, therefore, are likely to be hereditary in other breeds with high prevalence.<sup>2,3,9,10</sup> Heredity is also considered a risk factor for an intrahepatic vascular anomaly that caused development of multiple extrahepatic shunts in a family of Cocker Spaniels.<sup>11</sup> Besides heredity, no other risk factors for development of CPSS have been reported in dogs, although results of 1 study<sup>5</sup> suggests that puppies from small litters are more likely to have CPSS.

In our study, annual diagnostic proportion of CPSS in all dogs registered in the VMDB increased during a 21-year period from 1 of every 2,000 accessions to 1 of every 200 accessions. Diagnosis of CPSS in Yorkshire Terriers was a significant factor in this increase. Increasing annual diagnostic proportion of CPSS in Yorkshire Terriers may be caused by increased intensity of inbreeding, increased awareness of the disease by veterinarians and breeders, availability of tests for diagnosis of the disease, or a combination of these factors. The first major report<sup>12</sup> of CPSS in dogs was published in 1974, and numerous studies, reviews, and case reports have subsequently been published. However, the proportion of diagnoses of CPSS in dogs has not increased at a constant rate in all breeds. Annual prevalence of CPSS in Yorkshire Terriers increased 12-fold from 0.6% in 1980 to 7.2% in 2000, whereas the annual prevalence in all other dog breeds combined increased 6-fold from 0.04% to 0.24% in the same time period.<sup>2</sup>

In other studies,<sup>6,7,13-20</sup> Yorkshire Terriers, Miniature Schnauzers, Cairn Terriers, Maltese, Golden and Labrador Retrievers, Old English Sheepdogs, Irish Wolfhounds, German Shepherd Dogs, Poodles, Dachshunds, Pugs, Shih Tzus, Australian Cattle Dogs, and mixed-breed dogs have been reported with CPSS more frequently than other breeds. Golden and Labrador Retrievers, Australian Cattle

Dogs, Old English Sheepdogs, and Maltese were observed to be overrepresented, compared with the hospital population, although thorough statistical comparisons were not performed.<sup>6,20</sup> In our study, Australian Cattle Dogs, German Shepherd Dogs, and Dachshunds did not have significantly increased odds of CPSS, compared with the reference population. One reason for this discrepancy may be that these reports did not evaluate the proportion of accessions of CPSS among breeds and results for each breed were not compared with a standard or reference population.

The proportion of diagnoses of CPSS appears to vary with geographic location. Australian Cattle Dogs bred in Australia may have a greater prevalence of CPSS, compared with those bred in North America.<sup>7,20</sup> This may be caused by environmental or genetic characteristics of the regional population, or simply a greater awareness of CPSS in Australian Cattle Dogs among veterinarians and owners in Australia. Because Australian Cattle Dogs in the United States originated from dogs imported from Australia, the genetic characteristics of the American and Australian subpopulations may not be that much different.<sup>7</sup>

In our study, Skye Terriers (1.2%), longhaired Chihuahuas (0.9%), Scottish Deerhounds (0.9%), and Lakeland Terriers (0.6%) were 4 of the 15 breeds with the greatest proportions of diagnoses of CPSS during a 22.2-year period. However, the risk of CPSS for those breeds was not significantly greater than that of the reference population. The ability to detect a substantial increase in proportion of diagnoses of CPSS in individual breeds of dogs is affected by the total number of accessions of dogs of that breed. All 4 of these breeds had < 350 accessions. When the sample size is small, the adjusted confidence interval for the ORs can be sufficiently large to preclude a finding of significance. Further study of rare breeds is necessary to provide evidence for a risk of CPSS.

Prevalence is best determined by identifying animals with a disease in a defined population at a single point or during a specified period of time. In 1 study,<sup>5</sup> all Irish Wolfhound puppies born in the Netherlands from January 1, 1992 to January 1, 1997 were tested for CPSS at 2 to 3 months of age and the prevalence of the disease was 2.3%. In our study, the proportion of diagnoses of CPSS in Irish Wolfhounds in the VMDB population was lower (0.5%). Although this may be because of environmental or genetic differences in Irish Wolfhounds, the prevalence of CPSS in our study may also have been underestimated because diagnostic tests may have been limited to those suspected of having CPSS and that had owners who had the resources to pay for testing and treatment, dogs that died at birth or before referral were not included, and affected dogs may have been evaluated at primary care facilities without referral to a veterinary teaching hospital included in the VMDB.

Dogs reported in the VMDB are a subset of the population and do not accurately represent the population of dogs evaluated in primary care facilities or those in the general population. Dogs may be selectively referred to veterinary teaching hospitals for CPSS, compared with other diseases, because of the availability of surgi-

cal expertise. Although the potential exists for the effect of selection to be different among breeds, we believe that this was less likely. Once a diagnosis of CPSS is made or suspected in the primary care facility, we believe that it is more likely that economics or the attitude of the client toward referral, rather than breed of dog, will have a greater effect on whether the dog is evaluated at a veterinary teaching hospital. These factors would be expected to affect all breeds equally. Therefore, comparisons of the proportions of diagnoses of CPSS used as a surrogate for differences in prevalence of CPSS among breeds are likely to be valid. Populations of dogs accessed by veterinary teaching hospitals may include a greater percentage of dogs with diseases, compared with a primary care facility. Therefore, the true prevalence of CPSS within these breeds may be lower. Increases in the proportion of diagnoses of CPSS in the VMDB population with time may reflect trends in the general population or increase awareness of CPSS and referral of affected dogs.

In our study, we identified certain breeds that may be at increased risk for CPSS. Breeders should be made aware of the increased risk of CPSS in those breeds, and veterinarians should educate owners to look for clinical signs of the disease. Pre- and postprandial bile acids or ammonia tolerance tests should be recommended for puppies from breeds with increased risk of CPSS to determine whether liver dysfunction can be detected and further diagnostic tests should be pursued. Dogs with CPSS from breeds at increased risk for the disease should be neutered to prevent perpetuation of the disease. Advances in genetic testing may permit breeders to selectively reduce the risk of CPSS in their dog populations.

<sup>a</sup>Veterinary Medical Database, School of Veterinary Medicine, Purdue University, West Lafayette, Ind.

<sup>b</sup>SAS, version 8.02, SAS Institute Inc, Cary, NC.

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