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## Inherited defects in pedigree dogs. Part 1: Disorders related to breed standards

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## ABSTRACT

The United Kingdom pedigree-dog industry has faced criticism because certain aspects of dog conformation stipulated in the UK Kennel Club breed standards have a detrimental impact on dog welfare. A review of conformation-related disorders was carried out in the top 50 UK Kennel Club registered breeds using systematic searches of existing information. A novel index to score severity of disorders along a single scale was also developed and used to conduct statistical analyses to determine the factors affecting reported breed predisposition to defects. According to the literature searched, each of the top 50 breeds was found to have at least one aspect of its conformation predisposing it to a disorder; and 84 disorders were either directly or indirectly associated with conformation. The Miniature poodle, Bulldog, Pug and Basset hound had most associations with conformation-related disorders. Further research on prevalence and severity is required to assess the impact of different disorders on the welfare of affected breeds.

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## Introduction

Appearance matters in the pedigree-dog industry. Pedigree dogs are selected to conform to breed guidelines that are published but nevertheless open to interpretation and review. Dogs that best meet their breed's standards are rewarded in the show ring. Recently the United Kingdom pedigree-dog industry faced much criticism because certain aspects of dog conformation stipulated in the UK Kennel Club (KC) breed standards can have a detrimental impact on the dog's health and welfare (McGreevy, 2007; Higgins and Nicholas, 2008). The purpose of this review is to examine the number, prevalence and impact of disorders related to conformation aspects of the breed standards. This review is part of a larger study into all inherited disorders in pedigree dogs. The second part of this work deals with non-conformational inherited disorders (Summers et al., 2009).

Conformational breed-associated defects were recognised as early as 1868 by Charles Darwin, who hypothesised that muscular defects in Scottish deerhounds were related to their great size. In 1963, a review conducted by the British Small Animal Veterinary Association (BSAVA) at the request of the KC identified 13 abnormalities and defects in pedigree dogs, namely, hip dysplasia, patella luxation, entropion, retinal atrophy, elongated soft palate, abnormal temperament, skin-fold dermatitis, uterine inertia, elbow dysplasia, lens luxation, ectropion, trichiasis and deafness (Hodgman, 1963). At least 10 of these defects are associated with

conformation in some way. For example, an elongated soft palate is associated with a shortened muzzle and entropion is linked with skin folds around the eye.

In addition to phenotypic changes, contemporary modifications to the dog's environment may have resulted in an increase in the expression of certain breed-associated defects. For example, with advancements in veterinary therapeutics, animals tend to live longer and some conditions that manifest only in older age are more evident. Advances in veterinary science mean that more disorders can be detected, diagnosed and treated. For defects that can be corrected at an individual level by routinely available procedures, such as adnexal surgery in Shar Pei puppies, there is less incentive to address the occurrence at population level. In combination, the improved quality and availability of dog food and growing acceptance of the dog as a family member have contributed to a canine obesity epidemic (Slupe et al., 2008). Heart defects, musculoskeletal problems and endocrinological problems can all be exacerbated or become symptomatic through inappropriate feeding.

To date, research has primarily considered inherited disorders in isolation. However, to be able to make informed decisions about the impact on dog welfare of conforming to breed standards, inter-disorder comparisons are necessary to bring together clinical and epidemiological research. There is currently no standardised method of comparing the impact of different disorders on the health and welfare of dogs along a single scale. Quality of life (QoL) scales exist in the human medical literature (see, for example, Horn, 1983; Young-Saleme and Prevatt, 2001) and are used to grade the severity of a disease or injury to an individual in order to inform treatment options and allow evaluation and monitoring of health care services (Fine et al., 1995).

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Scoring systems can be disease- or organ system-specific to assess diagnostic groups or patient populations, or they can be generic, being applicable across impairments, illnesses and patient populations (Patrick and Deyo, 1989). The score is typically generated by categorisation of the illness according to features that are observable or can be measured. Individual-based non-generic QoL scoring systems are beginning to emerge in veterinary research (Wiseman-Orr et al., 2004). However, for the purposes of this research it was necessary to be able to compare disorders at the disorder- rather than the individual-level. Therefore, a secondary aim of the review was to develop a generic illness severity index for dogs (GISID) that could be applied to each disorder identified.

## Materials and methods

Due to the scale of pedigree-dog breeding as a whole, our review focussed on the top 50 KC-registered breeds in the UK compiled from KC registration statistics for 2007 (Kennel Club, 2008). In addition, data from KC registrations from 1998 to 2007 allowed us to estimate recent UK pedigree-dog population dynamics.

### Literature search

A wide variety of published scientific, policy, breeding and veterinary resources was used, including scientific literature, conference proceedings and veterinary textbooks, in addition to all breed club websites for the top 50 breeds, and international KC websites (for a complete list, see electronic Supplementary material).

Three existing online databases of inherited disorders in dogs (List of Inherited Disorders in Animals, LIDA<sup>1</sup>; Canine Inherited Disorders Database, CIDD<sup>2</sup>; and Inherited Diseases in Dogs, IDID<sup>3</sup>) were searched to produce a comprehensive list of all inherited disorders reported in the top 50 most popular breeds. A breed was considered to be predisposed to a condition if (1) the literature searched contained evidence of an increased incidence or prevalence in a breed, or (2) the literature searched made reference to a breed disposition or suggested in any terms that a breed had an increased susceptibility. This latter category was necessary due to the limited prevalence and incidence information available on this subject. However, the data collected was unfiltered meaning there will be a spectrum of robustness in the information reviewed here.

Using this comprehensive list of inherited disorders, we conducted a systematic search of the scientific and veterinary literature, using online bibliographic databases such as PubMed, Web of Knowledge and Google Scholar, employing the following search criteria:

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[Breed name] AND [Disease name].
[DOG OR CANINE] AND [Disease name].
[Breed name].
[INHERITED] AND [DOG OR CANINE].
[CONGENITAL] AND [DOG OR CANINE].
[GENETIC] AND [DOG OR CANINE].
[Disease name].
[INHERITED] AND [Breed name].
[CONGENITAL] AND [Breed name].
[GENETIC] AND [Breed name].
```

From this literature trawl we excluded case reports, studies conducted on research animals, and literature not published in the English language. In addition to the areas outlined above we extracted information from the publications concerning (1) the primary organ system affected; (2) details relevant to the prognosis, treatment, complications or impact on behaviour; (3) prevalence; (4) age of onset; (5) links with conformation; (6) related disorders, and (7) any other information of potential relevance to the study. Prevalence data extracted from the published literature included some screening systems or registries, including the Canine Eye Registry Foundation (Genetics Committee of the American College of Veterinary Ophthalmologists, 2007), the Orthopaedic Foundation for Animals (OFA)<sup>4</sup> and the BVA/KC hip dysplasia scheme.

### Severity score

A severity scoring system was developed to provide inter-disorder comparisons using information available from the scientific literature (Fig. 1). The scale was based on similar severity indices from human medicine (Horn, 1983; Young-Saleme

and Prevatt, 2001) in consultation with veterinarians and animal welfare scientists (non-veterinary researchers with expertise in measuring the behavioural and physiological consequences of animal welfare).

The prognosis score was developed to reflect temporal aspects of the disorder (i.e., whether the disease was chronic or acute), to allow for the possibility of lasting impairment and that the disorder might be fatal (either directly or indirectly through euthanasia). The treatment aspect of the score incorporates medical, surgical and side effects of treatment. Equivalence between medical and surgical therapies was based on assumed recommendations of such available treatments by veterinarians to clients. For example, we assumed that a clinician would recommend equally repeated minor (not intra-cavity) surgery and medium-term medical treatment that manages a disorder, but would favour these options over deep, intra-cavity surgery. Complications were included to represent the impact these could have on health and wellbeing, and behaviour was included to reflect QoL.

Each aspect was scored on a five-point scale from 0 to 4 with 0 being the least severe and 4 being the most severe. The four aspects of the index were summed to give a minimum total score of 0 and a maximum of 16. When scoring disorders we assumed that disorders were being treated in the most appropriate manner and that the environment and care provided were of the highest quality. We allowed for ranges of severity within diseases by scoring each disorder in its mildest and most extremes forms.

### Categorising disorders

Disorders were classified as conformation-related (C) if the disorder was reported to result directly from selection for a conformational trait; as a conformation-inherited link (CD) if the disorder was reported to be an inherited disorder exacerbated by a conformational trait; or as a non-conformational disorder (D) if the disorder was an inherited disease that showed no link with conformation in the literature reviewed. Classification as C, CD or D was based on expert opinion as extracted from the published literature. We categorised conditions according to the body system they primarily affected and C and CD disorders were tagged according to the aspect(s) of the KC breed standards that contributed to the condition, and according to skull morphology. Breeds were considered brachycephalic (high width:length ratio), mesocephalic (medium width:length ratio) or dolichocephalic (low width:length ratio) according to classifications found in the literature. For a full explanation of skull morphological classifications see electronic supplementary material.

This paper considers only C and CD disorders; D disorders are detailed in Summers et al. (2009).

### Statistical analysis

Disorders were totalled for breeds and body systems. For each breed we summed the severity scores of disorders to which that breed was predisposed. Non-parametric statistical tests were conducted to look for factors that affected the number of disorders associated with a breed, where each breed was considered as an independent data point. Spearman's correlations tested for associations between number of disorders per breed and popularity (number of KC registrations 2007), increase in popularity (change in KC registrations 1998–2007), breed height (median of range specified in UK KC Breed standards 2007), and breed weight (median of range specified in UK KC Breed standards 2008 or Grandjean, 2003).

Differences between the number of disorders of brachycephalic, mesocephalic or dolichocephalic breeds were analysed using a Kruskal Wallis test. In addition to written description, relational frequencies between aspects of the breed standards and disorders in the different body systems were used to compile network-style graphs.

## Results

### UK top 50 KC-registered breeds

The most popular pedigree breeds in the UK were Labrador retriever (45,079 registrations), Cocker spaniel (20,883), English Springer spaniel (14,702), Staffordshire bull terrier (12,167) and German shepherd dog (12,116) (Table 1 and electronic Supplementary material). Overall, the number of KC registered dogs increased by 5.8% between 1998 and 2007. No information was available on the total number of dogs, or total number of registered and unregistered pure-breed dogs in the UK, so the increase in KC registrations over the 10 years between 1998 and 2007 may represent either an increasing trend to register pure-breed dogs, or an increase in the total population of dogs in the UK, or a combination of these factors.

The breeds that have undergone the greatest relative increases in popularity since 1998 are the Dogue de Bordeaux (up 1204.1%),

<sup>1</sup> <http://www.vetsci.usyd.edu.au/lida/>. Accessed October 2008.

<sup>2</sup> <http://www.upei.ca/cidd/intro.htm>. Accessed October 2008.

<sup>3</sup> <http://www.vet.cam.ac.uk/idid/>. Accessed October 2008.

<sup>4</sup> <http://www.offa.org>. Accessed November 2008.

## Generic index severity index for dogs (GISID)

### Prognosis

SHORT ISOLATED BOUT & COMPLETE RETURN TO NORMAL	MEDIUM LENGTH ISOLATED BOUT OR SUCCESSIVE SHORT BOUTS & RETURN TO NORMAL	EXTENDED BOUT & RETURN TO NORMAL OR SUCCESSIVE SHORT BOUTS AND MINOR LONG-TERM IMPAIRMENTS	UNREMITTING OR CHRONIC ILLNESS OR BOUT(S) WITH MAJOR LONG-TERM IMPAIRMENT	IMMINENT DEATH AS A DIRECT RESULT OF CONDITION OR CONDITION-RELATED EUTHANASIA
0	1	2	3	4

### Treatment

NONE REQUIRED OR NOT NECESSARY AS MINIMAL IMPACT ON HEALTH	<b>MEDICAL- IMMEDIATE CURATIVE &amp;/ OR SURGICAL- SINGLE CURATIVE MINOR* SURGERY</b> <b>SIDE EFFECTS- NONE OR VERY MINOR, SHORT-TERM</b>	<b>MEDICAL- SHORT TERM CURATIVE OR MEDIUM-TERM MANAGEABLE &amp;/OR SURGICAL- SINGLE CURATIVE INTRACAVITY SURGERY/REPEATED MINOR* SURGERY</b> <b>SIDE EFFECTS- MINOR</b>	<b>MEDICAL- LONG TERM CURATIVE OR LONG-TERM MANAGEABLE &amp;/OR SURGICAL- DEEP INTRACAVITY SURGERY</b> <b>SIDE EFFECTS - MANAGEABLE PAIN OR MODERATE</b>	NONE AVAILABLE OR <b>MEDICAL- PROLONGED PALLIATIVE TREATMENT &amp;/OR SURGICAL- MAJOR DEEP INTRA-CAVITY SURGERY</b> <b>SIDE EFFECTS- CHRONIC INTRACTABLE PAIN OR MAJOR</b>
0	1	2	3	4

### Complications

NO LINKED DISORDERS	PREDISPOSITION TO MINOR SECONDARY CONDITION	PREDISPOSITION TO MODERATE SECONDARY CONDITION	PREDISPOSITION TO MAJOR SECONDARY CONDITION	PREDISPOSITION TO CATASTROPHIC SECONDARY CONDITION
0	1	2	3	4

### Behaviour

•Maintenance •Ingestion		•Elimination •Social		•Locomotion
NONE OF ABOVE DISTURBED	ONE OF ABOVE DISTURBED	TWO OF ABOVE DISTURBED	THREE OF ABOVE DISTURBED	FOUR OR MORE OF ABOVE DISTURBED
0	1	2	3	4

**Fig. 1.** The generic illness severity index for dogs (GISID). Each aspect was scored on a five-point scale from 0 to 4 with 0 being the least severe and 4 being the most severe. The four aspects of the index were summed to give a minimum total score of zero and a maximum of 16. \* Minor surgery is defined as not intra-cavity.

Alaskan malamute (up 936.6%), and Pug (up 449.1%). Breeds with the greatest decreases in popularity since 1998 are the Yorkshire terrier (down by 54.0%), Rough collie (down 48.9%) and Dalmatian (down 45.8%) (Table 1).

#### C and CD disorders

In the top 50 breeds, a total of 396 inherited disorders were identified from the literature and other sources searched: there were 63 category C, 21 CD and 312 D disorders. In terms of the

body systems C and CD disorders primarily affected were as follows: 25 were musculoskeletal (20 Cs, 5 CDs), 20 integument (13 Cs, 7 CDs), 17 nervous-sensory (16 Cs, 1 CD), 7 cardiovascular (1 C, 6 CDs), 5 urogenital (4 Cs, 1 CD), 5 respiratory (all Cs), 3 gastrointestinal (all Cs), 1 immune (C), and 1 endocrine (CD). Prevalence estimates were found for 36 conditions, but of these, only 10 were specific to the UK population.

German shepherd dogs were reported to be predisposed to the greatest number of inherited disorders overall and the Dogue de Bordeaux had the least (Table 1). Miniature poodles were reported

**Table 1**

Registrations statistics, number of disorders (including conformational related disorders, C, conformation exacerbated disorders, CD, and disorders not previously linked to conformation, D) and cumulative severity of disorders by breed.

Breed	Total disorders (C + CD + D)	Cs	CDs	Cumulative severity range for Cs	Cumulative severity range for CDs	Popularity rank (number registrations)	Percentage increase in registrations
German shepherd dog	77	13	6	64–123	35–73	5 (12,116)	–42.18
Boxer	63	13	5	71–155	18–27	10 (8191)	–14.78
Poodle (miniature)	58	17	2	47–113	27–55	48 (1038)	–6.82
Retriever (Golden)	58	4	4	17–37	20–38	7 (9557)	–35.44
Spaniel (English Springer)	57	15	0	62–138	0–0	3 (14,702)	15.39
Retriever (Labrador)	55	9	2	29–79	10–19	1 (45,079)	25.3
Doberman	53	7	6	21–65	23–47	23 (2437)	–16.14
Poodle (toy)	51	14	3	64–134	25–41	33 (1671)	–3.35
Spaniel (Cocker)	51	11	0	36–96	0–0	2 (20,883)	47.93
Dachshund (miniature smooth/long haired)	50	12	4	55–117	19–35	26/42 (2112/1244)	77.93/–27.93
Great Dane	50	12	8	51–112	40–74	29 (1897)	–6.41
Beagle	45	5	2	18–45	6–10	25 (2124)	126.2
Miniature schnauzer	45	3	1	10–28	7–14	11 (5152)	101.33
Irish setter	44	6	5	41–64	18–42	50 (1029)	–28.69
Bulldog	42	16	2	50–150	9–18	41 (1258)	8.35
Basset hound	41	16	3	69–154	7–20	16 (3979)	97.76
Poodle (standard)	41	9	1	51–97	7–12	47 (1057)	–19.13
Shar Pei	38	14	1	46–125	2–6	27 (2040)	132.61
Collie rough	37	7	1	37–71	2–10	43 (1196)	–48.87
Dalmatian	36	9	2	39–86	13–17	34 (1657)	–45.81
West highland terrier	35	7	3	30–75	0–0	9 (8309)	–45.09
Shetland sheepdog	34	1	0	7–19	0–0	35 (1655)	–31.24
Chihuahua (long/smooth coat)	33	13	2	56–128	14–26	31/45 (1728/1143)	42.34/114.85
Pug	33	16	2	60–145	9–20	17 (3547)	449.07
German short-haired pointer	32	4	2	18–37	12–13	38 (1497)	5.57
Rottweiler	32	5	0	18–46	0–0	14 (4257)	–14.07
Weimaraner	31	5	1	14–38	9–13	20 (2724)	–0.69
Akita	26	7	0	33–70	0–0	40 (1375)	–11.12
Shih tzu	26	7	0	30–68	0–0	12 (51447)	21.05
Yorkshire terrier	26	10	1	47–97	7–14	15 (4055)	–54.01
Border collie	25	4	0	16–36	0–0	24 (2359)	5.08
Cairn terrier	25	3	1	11–28	7–14	30 (1873)	–18.46
Cavalier King Charles spaniel	25	10	1	51–102	7–12	6 (1422)	–10.08
Lhasa apso	24	9	0	40–90	0–0	13 (4713)	40.35
Scottish terrier	24	5	1	19–42	2–10	49 (1031)	–19.45
Alaskan malamute	23	3	0	13–28	0–0	44 (1161)	936.61
Bullmastiff	20	9	0	32–78	0–0	37 (1594)	–32.37
Bichon frise	17	3	1	12–28	7–14	21 (2694)	5.56
Bull terrier	17	4	2	12–32	11–19	18 (3335)	32.18
Border terrier	16	4	1	18–36	2–10	8 (8814)	153.35
Hungarian Vizsla	16	4	1	23–37	0–5	46 (1133)	114.58
Siberian husky	15	1	0	3–10	0–0	28 (2000)	151.26
Rhodesian ridgeback	14	5	0	26–53	0–0	36 (1618)	59.25
Tibetan terrier	13	3	0	19–36	0–0	39 (1384)	45.84
Staffordshire bull terrier	11	2	2	6–12	13–21	4 (12,167)	27.23
Whippet	10	5	0	8–27	0–0	19 (3043)	–28.69
Retriever (Flatcoated)	8	3	0	9–28	0–0	32 (1527)	18.19
Dogue de Bordeaux	4	1	0	1–9	0–0	22 (2543)	1204.1

to be predisposed to the most C disorders, followed by the Pug, Bulldog and Basset hound. Shetland sheepdogs, Siberian huskies, Staffordshire bull terriers and Dogue de Bordeaux were associated with the fewest C disorders. Great Danes, German shepherd dogs and Dobermans had the most CD disorders.

The number of C or CD disorders a breed was reportedly predisposed (in total and by system affected) was found to correlate with the percentage increase in registrations over the past decade, and the height and weight of the breed (Tables 2 and 3). Taller and heavier breeds had more associated C cardiovascular and gastrointestinal disorders and more CD integument and musculoskeletal disorders. Lighter breeds had more associated C respiratory and urogenital disorders and more CD endocrine disorders. Shorter breeds had more C nervous-sensory, respiratory and urogenital disorders. The numbers of CD disorders in the cardiovascular and

musculoskeletal systems were lower in breeds that have increased in popularity over the past decade.

Of the top 50 breeds 33 could be categorised as brachycephalic (14), mesocephalic (15) or dolichocephalic (4) (see electronic [Supplementary material](#)). References linking skull morphology with the remaining 17 breeds could not be sourced. Skull shape affected the respiratory disorders to which breeds were predisposed. Brachycephalic breeds had more respiratory disorders than mesocephalic and dolichocephalic breeds (Kruskal Wallis:  $Z = 9.75$ ,  $df = 2$ ,  $P = 0.008$ ).

A comprehensive table of disorders related to the top 50 breeds, their severity, prevalence, age of onset, relevant breed standard conformation and related disorders can be seen in the electronic [Supplementary material](#). The aspects of conformation that can lead to disorders are described below and Fig. 2a–f displays the breed

**Table 2**

Spearman's correlations between the number of conformational related disorders per breed and the number of registrations, the percentage increase in registrations (1998–2007), the height and weight of the breed (the median specified in KC breed standards and Grandjean, 2003). Statistically significant values are marked with \* if  $P < 0.05$  and \*\* if  $P < 0.01$ .

	Number of Kennel Club registrations	Percentage increase in registered dogs (1998–2007)	Height	Weight
Total	$\rho = 0.125$ , $P = 0.40$	$\rho = -0.190$ , $P = 0.20$	$\rho = -0.191$ , $P = 0.19$	$\rho = -0.051$ , $P = 0.73$
Cardiovascular	$\rho = 0.105$ , $P = 0.47$	$\rho = -0.188$ , $P = 0.20$	$\rho = 0.271$ , $P = 0.06$	$\rho = 0.290$ , $P = 0.05^*$
Gastrointestinal	$\rho = -0.068$ , $P = 0.65$	$\rho = -0.243$ , $P = 0.09$	$\rho = 0.369$ , $P = 0.01^*$	$\rho = 0.426$ , $P = 0.03^*$
Immune	$\rho = -0.184$ , $P = 0.21$	$\rho = -0.237$ , $P = 0.11$	$\rho = 0.068$ , $P = 0.64$	$\rho = 0.053$ , $P = 0.72$
Integument	$\rho = 0.236$ , $P = 0.11$	$\rho = -0.106$ , $P = 0.47$	$\rho = 0.068$ , $P = 0.65$	$\rho = 0.246$ , $P = 0.09$
Musculoskeletal	$\rho = 0.054$ , $P = 0.72$	$\rho = -0.062$ , $P = 0.68$	$\rho = -0.096$ , $P = 0.515$	$\rho = 0.017$ , $P = 0.91$
Nervous-sensory	$\rho = -0.028$ , $P = 0.85$	$\rho = -0.237$ , $P = 0.105$	$\rho = -0.300$ , $P = 0.04^*$	$\rho = -0.193$ , $P = 0.19$
Respiratory	$\rho = 0.263$ , $P = 0.07$	$\rho = 0.117$ , $P = 0.43$	$\rho = -0.409$ , $P = <0.01^{**}$	$\rho = -0.330$ , $P = 0.02^*$
Urogenital	$\rho = 0.188$ , $P = 0.20$	$\rho = 0.025$ , $P = 0.68$	$\rho = -0.515$ , $P < 0.01^{**}$	$\rho = -0.503$ , $P < 0.01^{**}$

**Table 3**

Spearman's correlations between the number of conformation exacerbated disorders per breed and the number of registrations, the percentage increase in registrations (1998–2007), the height and weight of the breed (the median specified in KC breed standards and Grandjean, 2003). Statistically significant values are marked with \* if  $P < 0.05$  and \*\* if  $P < 0.01$ .

	Number of Kennel Club registrations	Percentage increase in registered dogs (1998–2007)	Height	Weight
Total	$\rho = 0.039$ , $P = 0.79$	$\rho = -0.219$ , $P = 0.13$	$\rho = 0.034$ , $P = 0.81$	$\rho = 0.095$ , $P = 0.52$
Cardiovascular	$\rho = 0.158$ , $P = 0.28$	$\rho = -0.304$ , $P = 0.04^*$	$\rho = -0.013$ , $P = 0.928$	$\rho = -0.007$ , $P = 0.96$
Endocrine	$\rho = -0.016$ , $P = 0.91$	$\rho = -0.015$ , $P = 0.92$	$\rho = -0.248$ , $P = 0.09$	$\rho = -0.286$ , $P = 0.04^*$
Integument	$\rho = 0.001$ , $P = 0.998$	$\rho = -0.075$ , $P = 0.61$	$\rho = 0.260$ , $P = 0.08$	$\rho = 0.357$ , $P = 0.02^*$
Musculoskeletal	$\rho = -0.021$ , $P = 0.89$	$\rho = -0.322$ , $P = 0.03^*$	$\rho = 0.439$ , $P < 0.01^{**}$	$\rho = 0.386$ , $P < 0.01^{**}$
Nervous-sensory	$\rho = 0.205$ , $P = 0.16$	$\rho = -0.205$ , $P = 0.16$	$\rho = 0.132$ , $P = 0.37$	$\rho = 0.169$ , $P = 0.25$
Urogenital	$\rho = -0.183$ , $P = 0.21$	$\rho = 0.059$ , $P = 0.69$	$\rho = -0.047$ , $P = 0.75$	$\rho = -0.140$ , $P = 0.34$

standard conformations that most frequently lead to disorders in each body system.

### Conformation-related conditions

#### General appearance and characteristics

One general breed feature that is related to defects is the hair ridge peculiar to the Rhodesian ridgeback. The ridge is genetically linked to dermoid sinus (severity: 6–14) and recent estimates of the prevalence of this condition are between 5–6% in this breed

(Miller and Tobias, 2003; Hillbertz and Andersson, 2006). It has been suggested that if matings between ridged dogs were avoided this condition could virtually be eliminated (Hillbertz et al., 2007).

#### Head and skull, mouth, eyes and ears

Many conditions have been linked with the brachycephalic head shape, including stenotic nares, an elongated soft palate and hypoplastic trachea. Brachycephalic airway obstruction syndrome (BAOS) is a combination of these conditions with a wide range of severity (6–15). Breeds predisposed to this condition include Bulldogs and Pugs (Lorinson et al., 1997). The large head to pelvis ratio (found in certain brachycephalic breeds) has been linked with dystocia (severity: 2–6) (Tilley and Smith, 2004), as have extremes in size (Bergstrom et al., 2006). A recent study of Swedish breeds found incidence rates were highest in the Scottish Terrier, Chihuahua, Pug and Staffordshire Bull Terrier (Bergstrom et al., 2006).

Reduced cranial cavity size is associated with the potentially severe neurological conditions of cranioschisis (severity: 13–15), hydrocephalus (severity: 4–13) and syringomyelia (severity: 13–15). This last condition is most commonly associated with Cavalier King Charles spaniels. It is caused by a malformation of the skull, known as the Chiari-like malformation, and is supposedly related to selection for skull conformations that are steep caudally. The condition is assumed to be painful (Rusbridge, 2005), as well as causing brain damage, and long-term success of treatment is limited (Rusbridge, 2007).

Dogs with protruding or sunken eyes are prone to ulceration or irritation of the eye, including ulcerative keratitis, keratitis sicca and keratopathy syndrome. Such conditions are of concern despite their relatively low severity (scores range between 2 and 9) because of their high prevalence in certain breeds. For instance, of those screened by the American Canine Eye Registry Foundation (2007), nearly 8% of Shar Peis were found to have keratitis and 17% of Pugs had keratopathy syndrome.

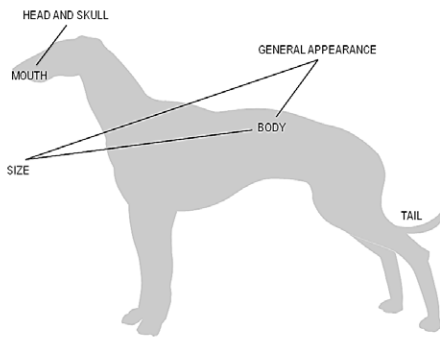
Otitis is frequently recurrent, often chronic in nature, painful (see, for example, Stern-Bertholtz et al., 2003), and can result in conductive deafness and aural tumours (Fan and de Lorimier, 2004; Kahn et al., 2005). Six of the top 50 breeds have a reported predisposition to otitis. Otitis externa has been linked with breed characteristics that predispose to skin infections and breeds with pendulous ears, excessive hair in and around the ear (Hayes Jr et al., 1987) and high production of cerumen, typically long-coated breeds (Strain, 1996).

#### Size, body, forequarters, hindquarters and tail

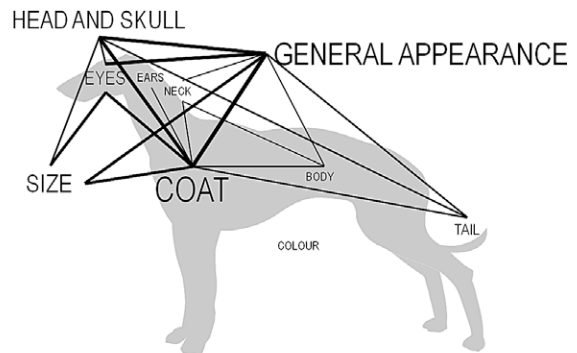
Large breeds may be predisposed to a number of disorders as a consequence either of their body size, or their fast rate of growth. Studies have demonstrated a high prevalence of hip and elbow dysplasias in many large and giant breeds (Sturaro et al., 2005; Genevois et al., 2008). Prevalence estimates have been as high as over 50% (UK) (Genevois et al., 2008). With such polygenic disorders, environmental factors are important. For example, appropriate diet and exercise have been shown to reduce the prevalence and severity, and delay the onset of hip (Sallander et al., 2006).

Conditions associated with small body size and particularly small leg size, include odontoid process dysplasia (severity: 7–14), shoulder dysplasia (severity: 4–7), and patellar luxation (severity: 6–9). Patellar luxation, which can cause lameness, reportedly primarily affects breeds from the Terrier, Toy and Utility groups. Prevalence data from the OFA (Footnote 4) for two of the worst affected breeds were 15% for Yorkshire terrier and 13.8% for Shar Pei. The condition is likely to be underreported because luxation may be transient (e.g., it occurs only during exercise).

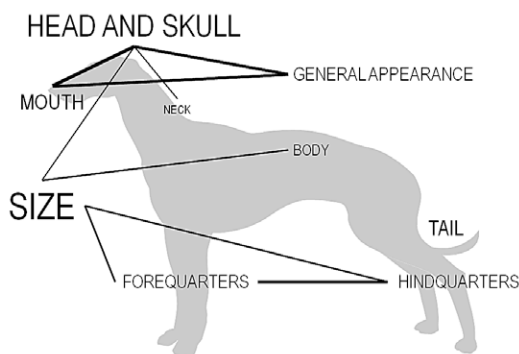
## (a) Gastrointestinal disorders



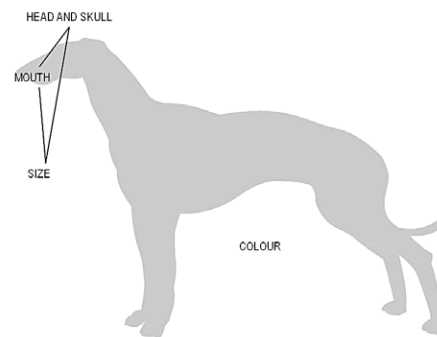
## (b) Integument



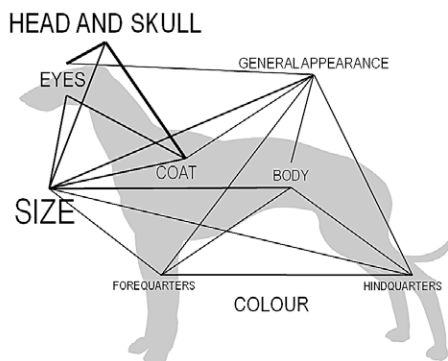
## (c) Musculoskeletal



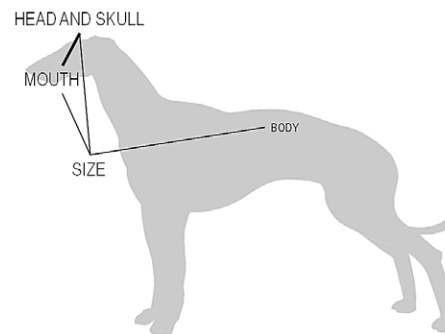
## (f) Urogenital



## (d) Nervous Sensory



## (e) Respiratory



**Fig. 2.** Aspects of the breed conformation that result in disorders in: (a) gastrointestinal, (b) integument, (c) musculoskeletal, (d) nervous-sensory, (e) respiratory and (f) urogenital systems. The size of the text indicates the number of disorders to which the conformational aspect has contributed. Interactions between conformational aspects are shown by the thickness of lines between aspects. Aspects were considered to interact when they contribute to the expression of the same disorder. The systems not shown (cardiovascular, endocrine and immune) had fewer than two disorders that resulted from conformation and these are detailed in the text alone.

Cervical vertebral instability and intervertebral disc disease are conditions that are both severe and of high prevalence in particular breeds. In a sample of 170 Dobermans in New Zealand, 49% were affected (Burbidge et al., 1994). Although evidence is mixed, cervical vertebral instability is linked with fast growth rates, heavy heads and long necks and has been reported in 12 of the top 50 breeds (Burbidge et al., 1994; Hedhammar et al., 1974). There are two types of cervical intervertebral disc disease. Type I is linked with chondrodystrophic breeds, such as Basset hounds and is caused by abnormal cartilage growth in the nucleus pulposus (McKee, 2000).

Conditions such as hemivertebrae (severity: 6–12) and spina bifida (severity: 5–11) are associated with selection for screw and curly tail shapes. A prevalence estimate for hemivertebrae could not be sourced, possibly because the condition has few clinical signs. In the dog population as a whole, the prevalence of spina bifida is 0.01% (Breit and Kunzel, 1998).

#### Coat and colour

Many disorders relate to wrinkled skin or excessive skin folds. Dermatitis and pyoderma may not be severe conditions, but they are often recurrent or chronic in nature (Hill et al., 2007). Although

a recent estimate could not be sourced, in 1963 skin-fold dermatitis had a high reported prevalence in Bulldogs (17% of 162 of this breed sampled). Skin-fold dermatitis has a high reported prevalence in Bulldogs (17% of 162 of this breed sampled; Hodgman, 1963), Cocker spaniels (7% of 318 sampled; Hodgman, 1963) and English Springer spaniels (17% of 24 sampled; Hodgman, 1963). Skin folds may be stipulated in breed standards directly or arise indirectly from the requirement for a brachycephalic skull shape, or corkscrew tails. The Shar Pei, a breed with many skin folds, presents with numerous skin complaints, including cutaneous mucinosis (severity: 3–7). It is proposed that selection for thickened wrinkled skin may have resulted in this condition since a large amount of dermal mucin is a cause of both cutaneous mucinosis and the thickened and wrinkled skin of the Shar Pei (Welle et al., 1999).

Entropion and ectropion are conditions of high prevalence in the Shar Pei (entropion, 58%; ectropion, 3%; CERF, 2007), Bulldog (entropion, 14%; ectropion, 11%; CERF, 2007), and Pug (entropion, 21%; CERF, 2007). In giant breeds, entropion and ectropion can co-occur such that the central lower lid is ectropic while at the corners the lid is entropic (producing diamond-shaped eyes). Conformational features that can affect the normal eyelid structure include a large distance between upper and lower lids, a small or recessed globe, a visible third eyelid or a drooping lower eyelid (van der Woerd, 2004). Two related conditions are trichiasis (severity: 2–9), which is found in breeds with nasal folds or droopy eyelids (Tilley and Smith, 2004), and eversion of nictitating membrane (severity: 1–5), which is found in larger breeds with facial folds and a distinctive stop (Martin and Gelatt, 2003).

The piebald, extreme white, and merle coat colourations and, often associated, hypopigmentation of the iris are all linked with a series of co-occurring nervous-sensory conditions, including sensorineural deafness (severity: 4–8), iris atrophy (severity: 6–12), and microphthalmia (severity: 3–12). It is proposed that deafness is linked to melanocyte numbers so that dogs with lighter eyes or coats are more likely to be deaf (Cattanach, 1999). Overall the prevalence estimates for microphthalmia are low, less than 2.5% even for reportedly predisposed breeds (CERF, 2007). However, this would be expected given the condition's association with colouration, which is often flexible within a breed standard. One breed where colour is specified is the Dalmatian, and Dalmatians are known to have high prevalence of deafness. One study of 22,873 Dalmatians in the UK found 18.4% were either bilaterally or unilaterally deaf (Wood and Lakhani, 1997). Another condition reported to be related to colouration is urate urolithiasis (severity: 6–8). A lack of white hairs within Dalmatian spots is associated with high acid excretion (Trimble and Keeler, 1938).

Cyclic neutropenia is a severe condition linked with colour dilution and found primarily in grey collies (Horwitz et al., 2004). Breed standards for the Rough collie select against grey colouration, stating that a slate colour is undesirable.

#### *Inherited disorders indirectly linked to conformation*

##### *Head and skull, mouth, eyes and ears*

Uterine inertia, a condition of unknown prevalence, ranges in severity and is linked with dystocia. It is exaggerated in breeds with heads that are comparatively large for their body size, such as Bull terriers and Scottish terriers, since more uterine force is required to expel the puppies.

##### *Size, body, forequarters, hindquarters and tail*

Large and giant breeds are believed to be more susceptible to cardiovascular conditions such as aortic stenosis (severity: 5–13), atrial septal defect (severity: 0–12), dilated cardiomyopathy (severity: 11), and tricuspid valve dysplasia (severity: 9–13). Some large breeds are also reportedly predisposed to valvular disease

(e.g. German Shepherd Dog), however, mitral valve disease (severity: 7–12) is more commonly associated with small dog breeds (Borgarelli et al., 2004), particularly the Cavalier King Charles spaniel. The prevalence estimates for mitral valve disease in the Cavalier King Charles spaniel range between 11–45%<sup>5</sup> (Hyun, 2005; Haggstrom et al., 1992; Darke, 1987).

The demanding growth period of larger breeds and associated overproduction of fibrous or calcareous matter may be linked to disorders, including zinc-responsive dermatosis (severity: 5–9), degenerative myelopathy (severity: 13–14), metabolic bone diseases (e.g., calcinosis circumscripta, osteosarcoma and polyostotic fibrous dysplasia), and immune disorders (e.g., masticatory muscle myositis and panosteitis). Careful dietary control during the growth period can help reduce the incidence of many of these disorders.

#### *Coat and colour*

Breeds with a long, heavy or dense coat are predisposed to certain skin conditions, such as acute moist dermatitis (severity: 1–6) if the coat is not adequately maintained. Some breeds, which have a breed disposition to skin disorders, can also easily develop pododermatitis (severity: 2–10), where the footpads become inflamed and sore. Breeds that are particularly susceptible are those with interdigital hair that retains humidity and allows pyogenic proliferation.

Black hair follicular dysplasia (severity: 4) and colour dilution alopecia (severity: 4–9) are similar disorders linked to black or dark brown colouration and dilute colours such as fawn or blue colorations, respectively (Schmutz et al., 1998). It is believed that they are caused by irregular pigment clumping, which results in areas of weakness (Mecklenburg, 2006).

Another condition purportedly linked with colouration is demodicosis (severity: 2–6). Demodicosis, a cutaneous disease caused by the mite *Demodex canis*, is also more common in dogs with a red-coloured coat and short-haired dogs (Day, 1997).

## **Discussion**

According to the literature reviewed, every one of the 50 most popular pedigree-dog breeds has at least one aspect of its physical conformation that predisposes it to a disorder. In total, 84 disorders were either directly or indirectly associated with conformation. The number of disorders to which a breed was reported to be predisposed was related to size, with smaller breeds having more C or CD disorders affecting the nervous-sensory, respiratory, urogenital and endocrine systems and heavier breeds having more affecting the cardiovascular, gastrointestinal, integument and musculoskeletal systems. It was interesting to note that while no directional relationship was found between breed standards and C-type musculoskeletal disorders, this was not the case in CD disorders.

Heavier and taller breeds had more CD disorders, which are those exacerbated by a conformational trait. Large dogs may be particularly prone to musculoskeletal disorders because their bodies are at the upper threshold of the weight that can be supported. In combination with overfeeding or inappropriate feeding, the musculoskeletal system is pushed further beyond this threshold. A relationship between weight and certain musculoskeletal disorders, such as hip dysplasia, is established (Comhaire and Snaps, 2008). The link between brachycephalic head shape and respiratory disorders is also established (Lorinson et al., 1997). In contrast, the link between heavy breeds and disorders of the integument has not previously been reported. There are a number of ways this finding could be explained. For instance, the increased

<sup>5</sup> <http://www.cavalierhealth.org/>. Accessed November 2008.

volume of skin could cause defects related to the way the skin hangs, or could result from poorer maintenance due to the extra time required to brush a large dog.

Some findings from this study must be interpreted with caution. The literature searched was not limited to recent UK-based studies and was unfiltered. There will, therefore, be a range in the robustness of evidence used to determine supposed predispositions. Furthermore, this review is heavily biased towards reporting breed dispositions because it considers a breed predisposed if this is reported but does not remove this association if another publication finds a lack of association between the breed and disorder in question. This review has provided an appraisal of the current level of knowledge on the conformational-related disorders in pedigree dogs and enables identification of targets for future studies. It has also revealed patterns breed predisposition to conformational-related disorders.

### *Breeding to standard*

By selecting for appearance rather than function or health, many breeds have become predisposed to health problems (McGreedy and Nicholas, 1999). Some breed standards may be encouraging breeders to select for dogs predisposed to disease. One example of this is the Dalmatian. Although Dalmatians with patches are less likely to suffer from deafness (Henley and Wood, 2003), the breed standard states that 'patches are unacceptable'. Another example is the Pug. Breeds with screw-tails or curly tails are predisposed to spina bifida and hemivertebrae, but the breed standard for Pugs specifies that the tail should be '*curled as tightly as possible over hip, double curl highly desirable*' (Kennel Club, 2008). The Pug breed club<sup>6</sup> in the UK is aware of the high incidence of hemivertebrae in Pugs and has recommended that all dogs should be X-rayed before breeding. This move is laudable but will not eliminate the disorder if the breed standard is not altered.

Brachycephalic dogs are predisposed to elongated soft palates and stenotic nares, which lead to partial obstruction of the upper airways. Shortened faces are often accompanied by facial wrinkle, which can lead to multiple eye conditions. One breed standard – the Pekinese – has recently been changed to try to improve health and address such disorders and several other breed standards are currently under review by the Kennel Club.<sup>7</sup>

Certain conformational characteristics that predispose a breed to one condition may provide protection against another condition. For example, the OFA<sup>4</sup> statistics for elbow dysplasia show that 15 out of 20 breeds with the highest prevalence ranged in weight above 30 kg, but only one breed of this weight range is found in the 20 breeds with the highest prevalence of patella luxation.

### *Estimating the scale of the problem*

The establishment of a generic illness severity index for dogs (GISID) was a significant innovation that offers a framework for ethical and cost-effective decision-making in the care of dogs with inherited disorders and the debate around pedigree-dog breeding. Although in human medicine, severity scoring systems are common (Endicott et al., 1976; Young-Saleme and Prevatt, 2001) and researchers have spent many years developing and validating them, such a system does not appear to exist in the veterinary world. The severity index developed by the current study gives the range of severity of possible complications and behavioural changes associated with each disorder.

The score could be refined to take into account additional factors, such as the probability of complications, or the degree of impact on behaviour. However, this was not possible in the current study because such quantitative information is not provided in the literature and therefore our approach to score the presence or absence of complications or behavioural changes is justified. Scoring was at a disorder level, so the score included both the best and worst case scenarios for that disorder. For some disorders, this resulted in such a wide-ranging severity score (for instance patent ductus arteriosus scored 0–14) that the score loses its utility. To further refine the severity index, the range could be measured at the level of the individual or given as a probability distribution, showing the likelihood of disorders manifesting with different degrees of severity based on outcomes from a number of representative individuals. Such an approach would require targeted epidemiological studies of canine defects.

The initial searches for inherited disorders in dogs were conducted using the three online databases LIDA, CIDD and IDID. Each of these databases are freely available and contain a huge amount of information that can be used to inform the public, update veterinary clinicians, and inform research and policy. However, we found that no single database contained all the disorders associated with pedigree dogs, and that the databases used different terminology to refer to the same condition. In part, this could have resulted from the differences in the target audience of the database; LIDA and CIDD are designed for owners and potential owners, breeders and veterinarians, and IDID is primarily a resource for researchers. The specificity of the disease description and decisions such as whether to include complexes would have affected the number of disorders described, and the manner in which they were described. Other differences between the databases include the sources used to compile the database, the country of the data source(s) and the level of evidence required to describe a disease as inherited. LIDA, for instance, based classification of breed disposition on data from the USA whereas IDID considers any disorder which is '*likely to be transmitted wholly or partly through a genetic mechanism*'. LIDA and IDID contain links to scientific search engines, and thus can be used to locate the latest research on a particular disorder, but no database contains recent prevalence data.

Ideally, we would have been able to find prevalence data for the majority of the disorders. However, in the majority of cases, prevalence data were either completely lacking, not UK-based, not recent, or were drawn from biased samples. This is perhaps unsurprising since there are 4200 breed-disorder combinations, considering the top 50 pedigree breeds and the 84 C and CD disorders identified. Prevalence data is essential to assess the proportion of dogs affected by a condition and the degree to which a particular breed is prone to a particular condition. Such information is useful for clinical and policy-level decision making processes.

In this review, we have focussed on the top 50 breeds from the number of UK KC registrations, but it is likely that many pedigree and pure-bred dogs are not registered. This makes it very difficult to place numerical estimates on the proportion of dogs affected by inherited disorders even where prevalence estimates were available. With some notable exceptions (e.g., hip dysplasia; Comhaire and Snaps, 2008), we currently know very little about the frequency with which particular degrees of physical conformations result in, or exacerbate, particular disorders.

In combination with GISID, prevalence data would allow estimation of the welfare impact of different disorders on different breeds (for full method see Collins et al., submitted for publication). Whilst immediate action is clearly needed to safeguard the welfare of pedigree dogs, the long-term approach must include furthering our understanding of the relations between physical conformation and inherited disorders at the population-level.

<sup>6</sup> <http://www.pugdogclub.org.uk/start.html>. Accessed December 2008.

<sup>7</sup> <http://www.thekennelclub.org.uk>. Accessed December 2008.



## Conclusions

Whilst literature has linked each of the 50 most popular pedigree breeds of dog in the UK to at least one heritable defect due to physical conformation many of these links are not yet supported with accurate prevalence data. The association of some of these conditions with official breed standards and the high maintenance implications of some breed features make conformational extremes an area which needs to be addressed to safeguard the welfare of pedigree dogs in the future.

## Conflict of interest statement

This work was funded by the Dogs Trust but was commissioned to be an independent review. As such, none of the authors of this paper has a financial or personal relationship with other people or organisations that could inappropriately influence or bias the content of the paper.

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## Appendix A. Supplementary material

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.tvjl.2009.08.033.

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