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Welfare epidemiology as a tool to assess the welfare impact of inherited defects on the pedigree dog population

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Abstract

The effect that breed standards and selective breeding practices have on the welfare of pedigree dogs has recently come under scrutiny from both the general public and scientific community. Recent research has suggested that breeding for particular aesthetic traits, such as tightly curled tails, highly domed skulls and short muzzles predisposes dogs with these traits to certain inherited defects, such as spina bifida, syringomyelia and brachycephalic airway obstruction syndrome, respectively. Further to this, there is a very large number of inherited diseases that are not related to breed standards, which are thought to be prevalent, partly as a consequence of inbreeding and restricted breeding pools. Inherited diseases, whether linked to conformation or not, have varying impact on the individuals affected by them, and affect varying proportions of the pedigree dog population. Some diseases affect few breeds but are highly prevalent in predisposed breeds. Other diseases affect many breeds, but have low prevalence within each breed. In this paper, we discuss the use of risk analysis and severity diagrams as means of mapping the overall problem of inherited disorders in pedigree dogs and, more specifically, the welfare impact of specific diseases in particular breeds.

Keywords: *animal welfare, breeding, dog welfare, epidemiology, inherited disorders, risk analysis*

Introduction

Epidemiological methods are being used increasingly commonly in farm animal welfare research to understand the prevalence of a particular welfare problem, or welfare 'hazard', in a population and the risk factors associated with its occurrence. In addition to elucidating the prevalence of particular welfare hazards, quantifying the impact these hazards have on an individual animal's welfare is of increasing interest. This may involve quantifying duration of exposure to the hazard and the consequences of exposure, and the 'intensity' of the adverse effects of exposure (EFSA 2009). In farm animal welfare, the combination of prevalence and intensity is taken into account to permit a ranking of welfare problems according to the product of their severity (at the individual level) and how widespread they are in the population. Ultimately, rankings such as these could be useful in strategising the use of limited resources for welfare improvement, for example through legislative change, or the development of DNA markers for particular disorders. So far, these methods of risk analysis have been applied mainly in farm animal welfare contexts and have not been used to try to rank various welfare challenges in companion animals. In this paper, we apply these methods to quantify the welfare impact of inherited diseases in pedigree dogs.

Following a high profile BBC documentary aired in the United Kingdom in 2008, general awareness of the issues surrounding the welfare of pedigree dogs in relation to breed standards and selective breeding practices has been raised. Recent research has suggested that breeding for particular aesthetic traits, such as tightly curled tails, highly domed skulls and short muzzles predisposes dogs with these traits to certain inherited defects, such as spina bifida, syringomyelia and brachycephalic airway obstruction syndrome, respectively (Asher *et al* 2009; Rooney & Sargan 2009). These types of problems are neither new nor undocumented indeed, Charles Darwin wrote of his belief that the muscular defects observed in Scottish deerhounds were a direct consequence of their great size (Darwin 1868). Breed standards provide a written description of each pedigree breed to dictate the preferred configurations and conformations of the head, nose, eyes, ears, forequarters, hindquarters, body, tail, feet and the nature of the coat and colouration, and the relative dimensions of the dog (Kennel Club Breed Standards 2008). Asher *et al* (2009) found that every breed in the top 50 most popular breeds of dog in the

UK was predisposed to at least one inherited defect linked to breed standards and that, in total across these 50 breeds, there are 84 disorders directly or indirectly associated with specifications in the breed standards.

In addition to inherited defects associated with specific breed standards, there are a large number of disorders that are not related to breed standards but are inherited and thought to have emerged partly as a consequence of inbreeding and restricted breeding pools (McGreevy & Nicholas 1999). Recent work by Summers *et al* (2010) estimated a total of 312 inherited disorders of this type. How can we estimate the welfare impact of each of these diseases both individually and as a whole on the pedigree dog population? In this paper, we discuss the use of risk analysis and severity diagrams as means of mapping the overall problem of inherited disorders in pedigree dogs and, more specifically, the welfare impact of specific diseases in particular breeds.

Welfare epidemiology

Risk assessment is a systematic, objective process used to estimate the probability of exposure to a hazard and the consequences of that exposure. In risk assessment terms, a welfare hazard is any factor with the potential to compromise animal welfare. The risk associated with a particular hazard is a function of the likelihood of occurrence and the consequences of occurrence. In characterising a welfare hazard, three parameters are considered: (i) the intensity of the adverse effect that the hazard causes ('severity'); (ii) the duration of the effect; and (iii) the probability of exposure to the hazard (estimated from the prevalence). The product of these three (equally weighted) parameters gives us a welfare risk score (EFSA 2009).

Intensity or 'severity' of hazard

Asher *et al* (2009) and Summers *et al* (2010) assessed the severity of conformation-related and non-conformation-related inherited disorders affecting pedigree dogs using a novel severity index adapted from severity scoring systems used in human medicine. This index, the Generic Illness Severity Index for Dogs (GISID) and its development is described in Asher *et al* (2009). The GISID scores four factors — prognosis, treatment, complications and behaviour. Each factor is scored from zero to four, where zero is considered least severe and four most severe (Figure 1). So, the maximum possible GISID score is 16. The prognosis score is the typical expected duration (or range of durations) of the disease in a typical dog. Therefore, in this scoring system, duration was not considered separately from intensity, but was incorporated into the severity score under the prognosis domain (Figure 1).

Probability of hazard exposure

The probability of hazard exposure can be estimated using prevalence data, and in Asher *et al* (2009) and Summers *et al* (2010) a systematic review process was used to find prevalence estimates for each of the inherited disorders in the UK. Prevalence was recorded as the percentage of the population affected by a disorder at any one time. We focused specifically on prevalence in the UK as free public access to the UK Kennel Club breed registration statistics for the period in which we were interested (1998–2007) meant that we had an estimate of our population at risk. However, due to the lack of prevalence data in general, and in the UK specifically, for most of the inherited disorders to which pedigree dogs are exposed, the welfare impact of many of the disorders could not be estimated.

Welfare impact

Welfare impact (WI) is a function of the prevalence estimate for a particular welfare hazard within a specified population and the severity (or intensity) of that hazard at the individual level:

Welfare impact = Prevalence (%) × (GISID score/16)

The maximum possible welfare impact score is 100 — this would describe a population where every individual is exposed to a particular, catastrophic welfare hazard.

Where prevalence estimates were available, the welfare impact score was calculated (Table 1). According to the WI scores calculated, the inherited diseases with greatest welfare impact are collie eye anomaly (CEA) in Shetland sheepdogs (welfare impact [WI]: 27.0–54.0), rough collies (WI: 24.0–48.0) and Border collies (WI: 24.0–48.0); hip dysplasia in bull mastiffs (WI: 15.63–46.88), Newfoundland (WI: 15.63–46.88), German shepherd dogs (WI: 0.0–31.19) and golden retrievers (WI: 15.63–31.25); and entropion in bulldogs (WI: 7.5–33.75).

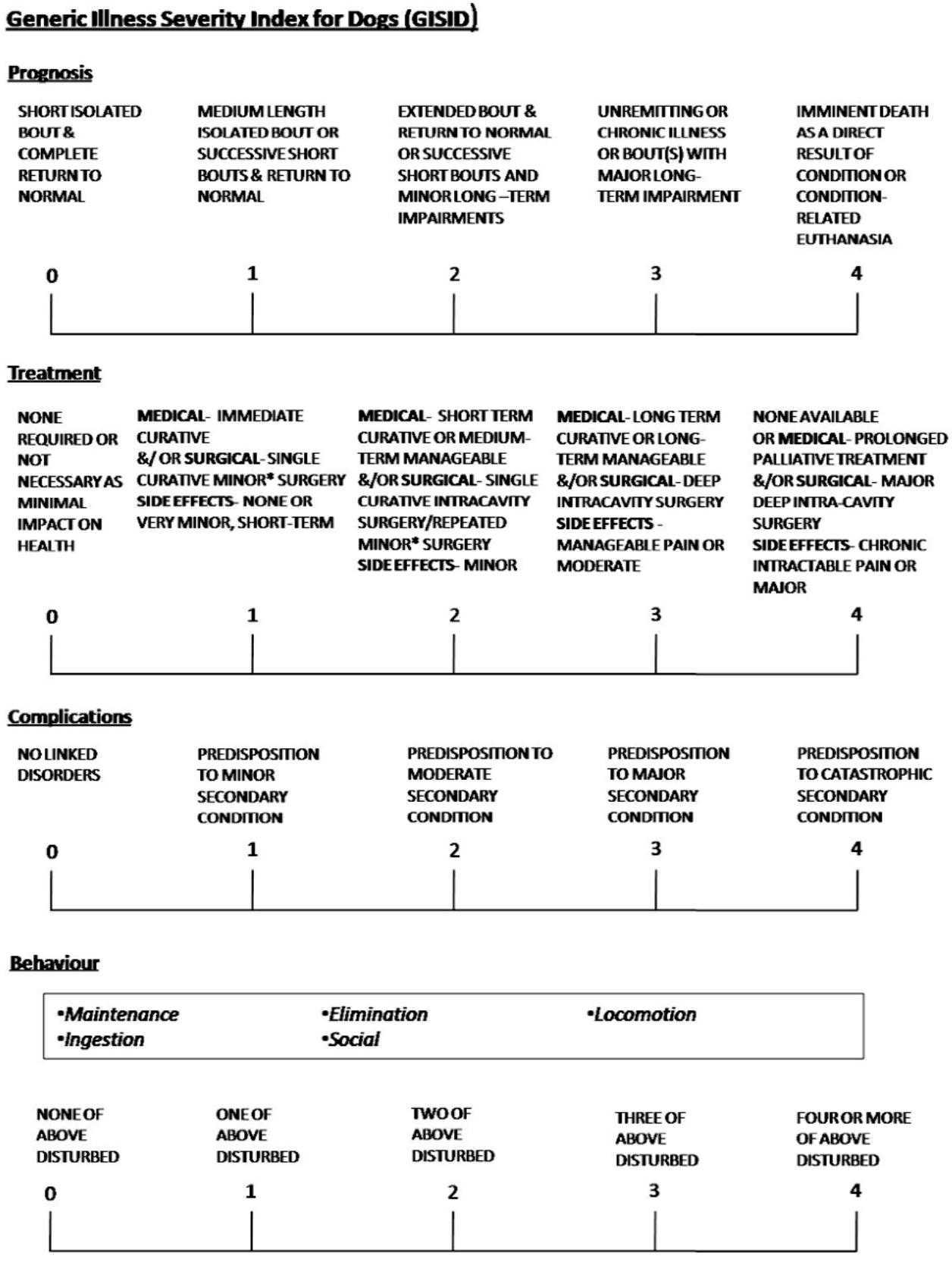
Comparing severity

Another method for comparing different inherited diseases in the absence of reliable prevalence data is to consider the relative severity of conditions. The GISID scoring system allows us to compare different diseases based on their intensity and duration, independently of prevalence information (see Figure 1, from Asher *et al* 2009). Kite diagrams can be used to compare the four factors between different diseases, and also to readily identify factors that differ most at the minimum and maximum overall severity levels. Here, we offer four examples of such comparisons.

Collie eye anomaly and glaucoma

Glaucoma and CEA are two nervous-sensory disorders affecting the eyes of the dog. CEA is a disease complex that involves a simple autosomal recessively inherited chorioretinal hypoplasia, caused by abnormal choroid development, with concurrent colobomatous defects (pits in or around the optic disk) (Bedford 1982). It affects predominantly collie-type breeds and is reported as being one of the most prevalent inherited eye disorders (Simpson *et al* 1998), though prevalence may be declining due to screening schemes for this and other conditions (KC/BVA/ISDS Eye Scheme, Schedule A, 2008 update). Glaucoma is a disease of the optic nerve and involves a loss of the retinal ganglion cells. It may be associated with high intraocular pressure. It is typically accompanied by acute pain and, if not treated promptly, a high risk of vision loss. There are numerous

Figure 1



The Generic Illness Severity Index for Dogs (GISID). Each aspect was scored on a five-point scale from 0–4 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. From Asher *et al* (2009).

Table 1 Welfare impact of inherited diseases in pedigree dogs in the UK.

Disorder	Prev (%)	GISID	Breed	Welfare impact	Welfare impact range	Reference
Dermoid sinus	18	6–14	Rhodesian ridgeback	6.75–15.75	9.00	1
Entropion	6	2–9	All	0.75–3.38	2.63	2
	60	2–9	Bulldog	7.50–33.75	26.25	2
	8	2–9	Bull terrier	1.00–4.50	3.50	2
	5	2–9	Boxer	0.63–2.81	2.18	2
	3	2–9	Cavalier King Charles spaniel	0.38–1.69	1.31	2
	3	2–9	Chihuahua	0.38–1.69	1.31	2
	1	2–9	Dalmation	0.13–0.56	0.43	2
	1	2–9	Toy poodle	0.13–0.56	0.43	2
	3	2–9	Miniature poodle	0.38–0.56	0.18	2
	25	2–9	Cocker spaniel	3.13–14.06	10.93	2
	4	2–9	English springer spaniel	0.50–2.25	1.75	2
	25	2–9	Cocker spaniel	3.13–14.06	10.93	2
	3	2–9	Golden retriever	0.38–1.69	1.31	2
	12	2–9	Labrador retriever	1.50–6.75	5.25	2
	2	2–9	Yorkshire terrier	0.25–1.13	0.88	2
Eversion of nictitating membrane	17	2–6	Bulldog	2.13–6.38	4.25	2
	7	2–6	Cocker spaniel	0.88–2.63	1.75	2
	17	2–6	English springer spaniel	2.13–6.38	4.25	2
Lens luxation	1	5–12	All	0.13–0.75	0.44	2
Trichiasis	1	2–9	All	0.13–0.56	0.43	3
	1	2–9	Cocker spaniel	0.13–0.56	0.43	3
	1	2–9	Boxer	0.13–0.56	0.43	3
Deafness	18.4	4–8	Dalmation	4.60–9.20	4.60	4
	4.5	4–8	Border collie	1.13–2.25	1.12	5
	36.3	4–8	Border collie (white head)	9.08–18.15	9.07	5
Aortic stenosis	33	5–13	Boxer	10.31–26.81	16.50	6
Dilated cardiomyopathy	1.4	11	All	0.96	0.00	7
Localised or juvenile onset demodicosis	0.01	2–6	All	0.00	0.00	8
	9.5	2–6	Bulldog	1.19–3.56	2.37	8
	6.3	2–6	Bull terrier	0.79–2.36	1.57	8
	1.7	2–6	Dobermann	0.21–0.64	0.43	8
	4.3	2–6	West Highland white terrier	0.54–1.61	1.07	8
Splenic torsion	0.01	6–12	All	0.00–0.01	0.01	9
Hip dysplasia	< 50	5–10	German shepherd dog	0.00–31.19	31.19	10
	50	5–10	Golden retriever	15.63–31.25	15.62	10
	25–40	5–10	Labrador retriever	7.81–25.00	17.19	10
	40	5–10	Bernese mountain dog	12.5–25.00	12.50	10
	20–25	5–10	Rottweiler	6.25–15.63	9.38	10
	> 50	5–10	Newfoundland	15.63–46.88	31.25	10
	< 20	5–10	Dobermann	0.00–12.44	12.44	10
	< 20	5–10	Irish setter	0.00–12.44	12.44	10
	< 20	5–10	Flat-coated retriever	0.00–12.44	12.44	10
	40	5–10	Boxer	12.5–25.00	12.50	10
	< 20	5–10	Bearded collie	0.00–12.44	12.44	10
	> 50	5–10	Bull mastiff	15.63–46.88	31.25	10
Collie eye anomaly	64	6–12	Border collie	24.00–48.00	24.00	11
	64	6–12	Rough collie	24.00–48.00	24.00	11
	72	6–12	Shetland sheepdog	27.00–54.00	27.00	11
Glaucoma	35	4–12	Flat-coated retriever	8.75–26.25	17.50	12

Welfare impact was calculated as the product of severity (based on the General Illness Severity Index for Dogs; GISID) and prevalence of the disease in the UK in the breeds stated in the table. Welfare impact is shown as a range, to reflect the variation in severity of each disease and is calculated by multiplying the minimum and maximum GISID scores (as a fraction of the maximum possible score, 16) by the prevalence. Impact range is calculated as the maximum welfare impact minus the minimum welfare impact and shows the amplitude of variation in welfare impact of a disease on a breed. References: 1) Mann & Stratton (1966); 2) Canine Eye Registry Foundation (2007); 3) Hodgman (1963); 4) Wood & Lakhani (1997); 5) Platt *et al* (2006); 6) Swift (1996); 7) Tidholm (1997); 8) Day (1997); 9) Neath (1997); 10) Coopman (2008); 11) Bedford (1982); 12) Wood *et al* (2003).

Figure 2

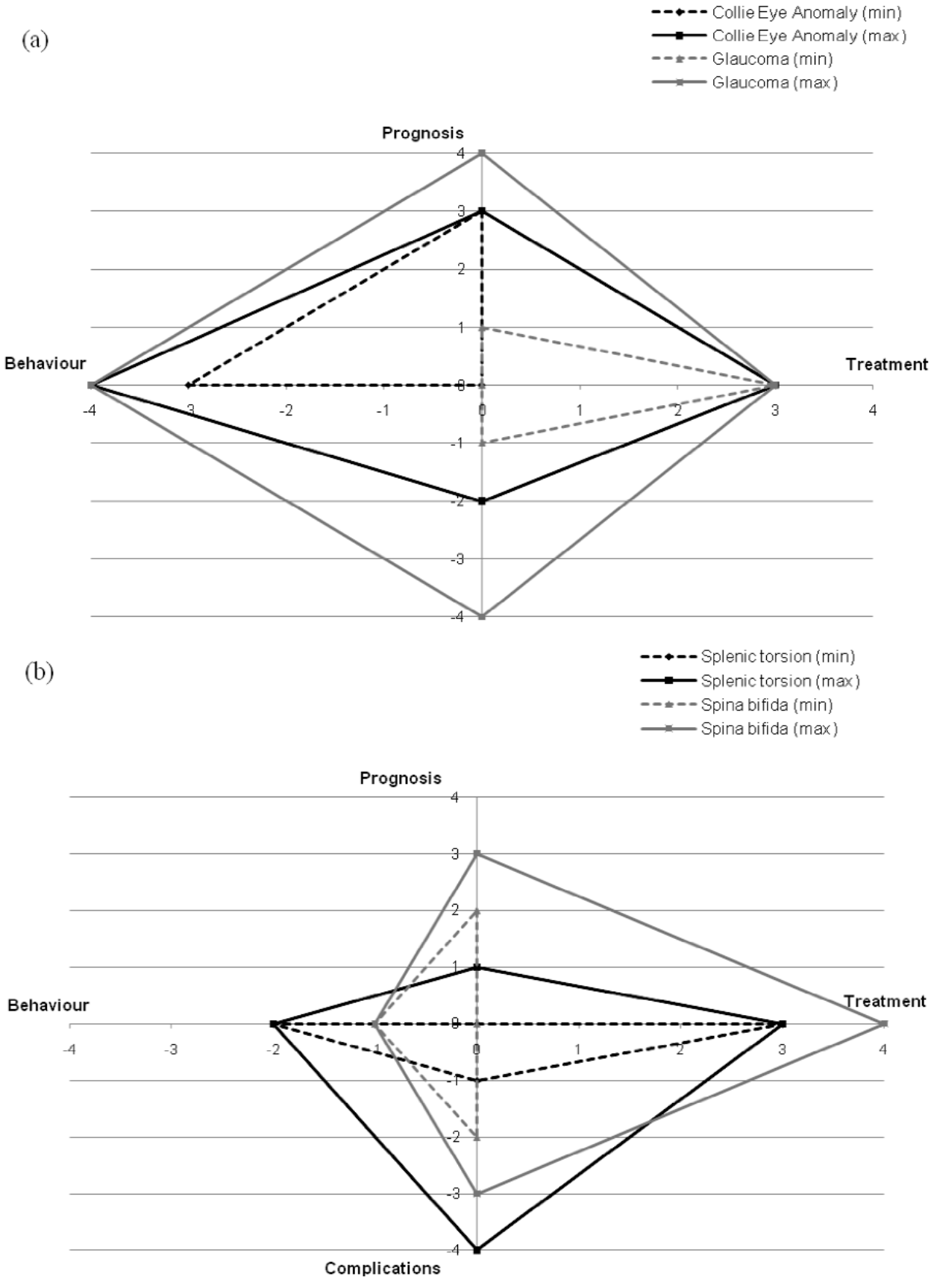
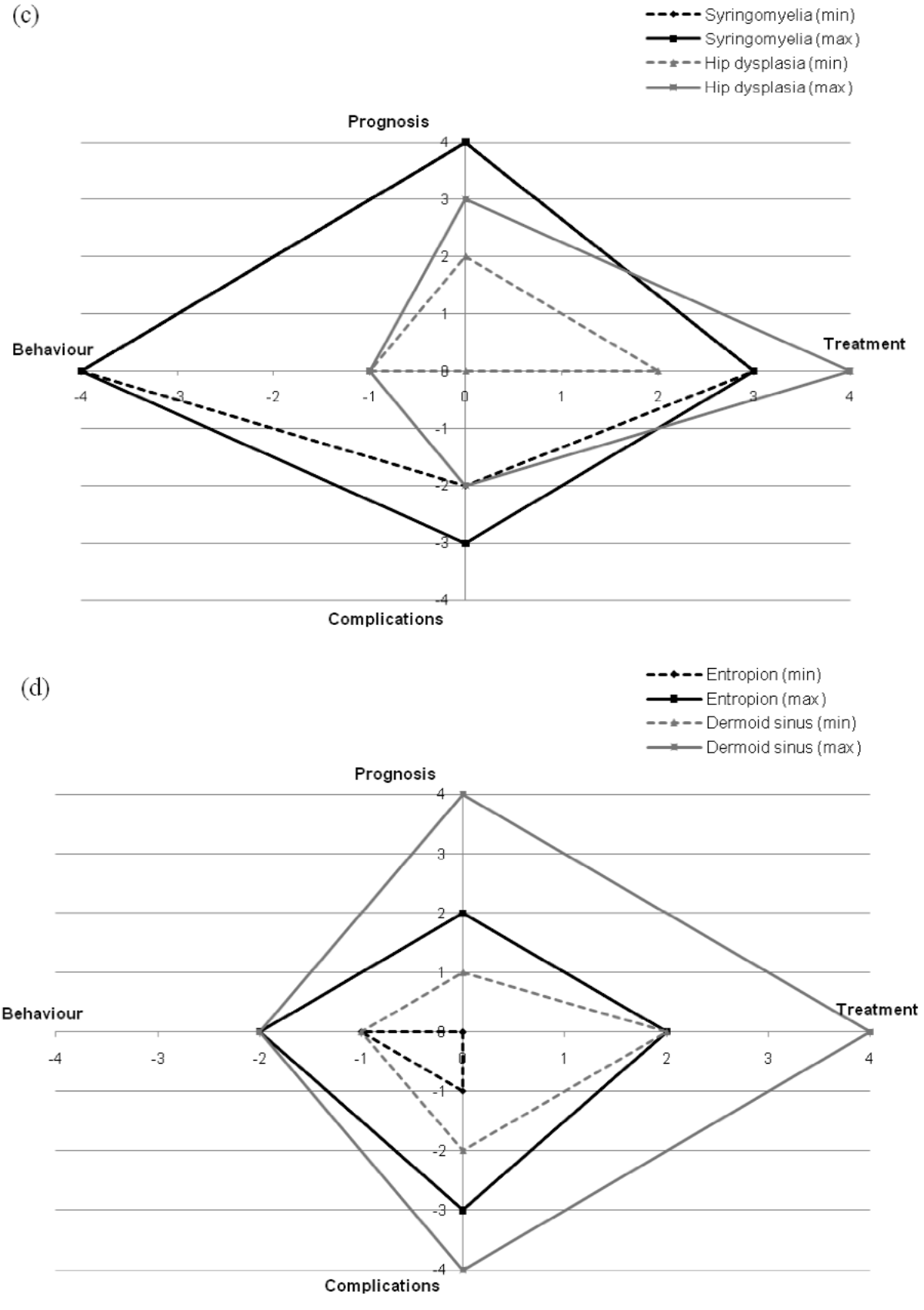

(a)
Collie Eye Anomaly (min)
Collie Eye Anomaly (max)
Glaucoma (min)
Glaucoma (max)
Prognosis
Behaviour
Treatment
4
3
2
1
0
-1
-2
-3
-4
-4
-3
-2
-1
0
1
2
3
4
(b)
Splenic torsion (min)
Splenic torsion (max)
Spina bifida (min)
Spina bifida (max)
Prognosis
Behaviour
Treatment
Complications

Figure 2 (cont)



Severity indices displayed as kite diagrams to compare the minimum and maximum severity scores of different inherited defects in pedigree dogs. For (a) Collie eye anomaly and glaucoma, (b) Splenic torsion and spina bifida, (c) Syringomyelia and hip dysplasia and (d) Entropion and dermoid sinus.

anatomical defects or conditions that are known or suspected to be predisposing factors for the development of glaucoma, though age of onset is often breed- and condition-dependent (Gelatt & MacKay 2004).

Figure 2(a) shows the severity kite range for CEA, according to the GISID scoring system. It highlights that the major difference between the least and most severe cases is primarily concerned with treatment. In the least severe cases, no treatment may be required, but in the most severe cases, major surgery may be required. However, if we compare CEA with glaucoma, we see that the factor that differs most between the least and most severe cases is behaviour. In the least severe cases, behaviour may be unaffected, but in the most severe cases, four or more basic types of behaviour (elimination, ingestion, locomotion, maintenance and social) may be impaired. There is also a difference in the level of possible complications and in the prognosis. However, in contrast to CEA, there is no difference in treatment.

Splenic torsion and spina bifida

Splenic torsion and spina bifida, though affecting different body systems, are thought to be associated with particular aspects of body conformation — the chest and the tail, respectively. Splenic torsion is categorised as a disease affecting the cardiovascular system (McGreevy *et al* 2005), in which the spleen rotates around its own axis and engorges with blood. Rotation can occur towards the stomach, potentially pulling the stomach with it, or away from the stomach, whereupon it can rotate multiple times. It is thought to affect 0.01% of the total dog population of the UK (Table 1), though primarily large, deep-chested breeds, such as Great Danes and German shepherd dogs are thought to be predisposed. Spina bifida is a musculoskeletal disease which is associated primarily with curly tailed breeds, such as the pug. It is thought that the selection for a tightly curled tail leads to twisting in the vertebral column, giving rise to a partially exposed neural tube.

Figure 2(b) shows the severity kites for both splenic torsion and spina bifida. Both disorders have a large range in possible severity but, again, the difference between the least and most severe cases is not seen in all four factors. The impact on behaviour is the same regardless of severity in both diseases. For spina bifida, the greatest difference is in treatment — in the least severe cases, no treatment is required, but in the most severe cases there may be no effective treatment available. The associated complications and prognosis of the disease change little between different cases. For splenic torsion, the greatest difference between the least and most severe cases is in the associated complications that can arise. These are the secondary diseases that an affected dog may acquire as a consequence of having splenic torsion, such as gastric dilatation-volvulus (gastric torsion, or bloat) and cardiovascular collapse, which can be far more severe than splenic torsion itself.

Hip dysplasia and syringomyelia

Hip dysplasia and syringomyelia are perhaps two of the most notorious inherited diseases in pedigree dogs due to their welfare consequences and high prevalence in certain breeds. Hip dysplasia is a malformation of the hip joint, whereby the femoral head only loosely or partially fits into the acetabulum and either one or both of these are misshapen, causing friction and wear within the joint. Hip dysplasia has traditionally been associated with larger breeds of dog — German shepherd dogs, Bernese mountain dogs, retrievers, Newfoundlands, bull mastiffs, boxers and rottweilers. Syringomyelia in Cavalier King Charles spaniels is considered to be a serious welfare issue as a consequence of its severity as a disease and its high prevalence in the population. Syringomyelia is caused by the development of cavitation in the spinal cord as a consequence of an obstruction of cerebrospinal fluid. It is thought to occur as a consequence of selection for domed skull shapes in the Cavalier King Charles spaniel breed, which causes crowding of nervous tissue at the posterior aspect of the head, leading to the development of Chiari-like malformation (Rusbridge 2005; Couturier *et al* 2008).

Figure 2(c) compares the severity kites for both hip dysplasia and syringomyelia. It shows the very narrow severity range for syringomyelia, illustrating that the only difference between the least and most severe cases is in the severity of the associated complications. The severity range for hip dysplasia is greater, though overall less severe than syringomyelia. The greatest difference between the least and most severe cases is in the treatment and possible associated complications, such as spondylosis, coxofemoral luxation and sciatica.

Dermoid sinus and entropion

The final comparison is between two diseases associated with the appearance of the integument. Dermoid sinus is a developmental condition found commonly in Rhodesian ridgebacks relating to the trichoglyphs (hair whorls) along the dorsum for which the breed is named. It occurs when the skin and the neural tube do not separate fully and manifests as a tube with an opening on the dorsal midline extending towards the vertebral column. Entropion is a disorder of the eyelid, where the eyelid turns into the eye, causing irritation. It is common in dogs with excessive sagging or, folds of skin, around the eyes.

Figure 2(d) compares the kites for dermoid sinus and entropion. In both cases, the difference between the minimum and maximum GISID scores is apparent in all four measured factors. Overall, entropion is a less severe condition than dermoid sinus and has less variability in the range of severity to which affected individuals are affected. In contrast, dermoid sinus can be potentially life-threatening, with some cases being so severe that no medical or surgical intervention is possible and that secondary complications are catastrophic.

Discussion

This paper has presented methods for quantifying and comparing the impact of inherited diseases, both at the individual and population level in pedigree dogs. At the population level, prevalence data and generic severity scores allocated using the GISID scoring system were used to calculate welfare impact scores for specific inherited diseases by breed. Using this method, the inherited diseases with the greatest welfare impact were collie eye anomaly, hip dysplasia (particularly in bull mastiffs, Newfoundlands, German shepherd dogs and golden retrievers) and entropion in bulldogs.

To investigate the combined impact of inherited diseases on all pedigree dogs in the UK, or compare the relative welfare impact of different diseases, we would need to take into account the different numbers of dogs of each breed. For example, a disease may be estimated to occur in 20% of Labrador retrievers and also in 20% of golden retrievers. The relative population sizes are significantly different — 45,079 Labrador retrievers were registered with the UK KC in 2007, compared with 9,557 golden retrievers (Kennel Club Registration Statistics 2007). Prevalence, therefore, needs to be weighted by population size. However, current population size estimates of different breeds are difficult to find because, although Kennel Club registers new dogs each year, there is no requirement for owners to notify the Kennel Club of the death of a registered dog.

Fluctuating population sizes, changes in prevalence and also changes to the GISID score (for example, through improved or new veterinary therapeutics) all affect the final welfare impact score. Consequently, each welfare impact score is a snapshot of the population at any one time and repeated assessments would be needed, to estimate how the impact is changing over time.

At the individual level, kite diagrams were used to visually compare the possible range in severity of different diseases, to highlight the differences in each of the four measured factors (prognosis, treatment, complications, behaviour) and show how these are linked to the least and most severe cases of the disease. Kite diagrams have previously been used in welfare assessment to compare the welfare of different species — laboratory animals, farm animals, companion animals and wildlife (Wolfensohn & Honess 2007). Applying this method usefully to different welfare problems would require the quantification or ordinal categorisation of each of the measured factors and also implies an equal weighting of each of these factors. The latter is rather more difficult to justify and indeed is one of the implicit assumptions of the GISID scoring system. Research is required to help to quantify the relative weighting of each of the four measured factors. Furthermore, in measuring the extent to which behaviour is affected, it is likely that loss of certain behavioural domains from the behavioural repertoire is more indicative of the severity of a condition than loss of other domains. The GISID system assumes, for example, that ingestive and maintenance behaviours are equivalent. However, dogs' motivation to perform maintenance behaviour may be rather lower than their motivation (when healthy) to feed, or drink water. If the baseline motivation levels for different behavioural domains differ, then the assumption that one behavioural domain is equivalent to another may be invalid and different behaviour types should be weighted according to baseline motivations in healthy dogs.

It should also be noted that kite diagrams do not show us the distributions of cases in terms of severity. Although we have plotted a range, for some diseases this range can be so wide, varying in severity from extremely minor to catastrophic, leading to imminent death. A consequence of this is that it becomes impossible to estimate the overall welfare impact of the disease on the dog population. For these diseases, among which are some considered to be of great importance to dog welfare, such as syringomyelia, dermoid sinus and collie eye anomaly, it is necessary to determine the exact shape of the severity distribution within the affected population. Currently, our assumption is that severity follows a uniform distribution, when a Gaussian curve may be more realistic. Further data is required in order to describe disorders in this manner, so that more accurate estimates of welfare impact can be generated for the pedigree dog population as a whole. In this way, decisions on how to tackle the problem of inherited disorders in pedigree dogs can be based on reliable and thorough information, and progress of any control or treatment methods employed can be successfully monitored over time.

Animal welfare implications

In the case of inherited diseases in pedigree dogs, there are approximately 400 diseases in the population as a whole. Understanding which of these to focus resources on requires an understanding of each of these disorders' effects at both individual and population level. The methods discussed in this paper could be potentially utilised for this purpose, although there is an immediate need for prevalence data and population information for this to be possible on a large scale.

Conclusion

Epidemiological methods, such as risk analysis, are established in farm animal welfare as a means of understanding risk factors associated with particular welfare hazards, and for prioritising where to focus resources for welfare improvement. Companion animal welfare challenges have not received the same level of risk analysis. This paper presented methods for quantifying and comparing the impact of inherited diseases both at the individual and population level in pedigree dogs. Individual-level comparisons were made between the least and most severe cases of different diseases using kite diagrams based on four scored factors which contribute to the severity of a disease (prognosis, treatment, complications and behaviour). These highlighted how diseases with similar severity scores can impact very differently on an individual. Calculating population-level welfare impact scores, we found that the inherited diseases with the greatest welfare impact were collie eye anomaly,

hip dysplasia (particularly in bull mastiffs, Newfoundlands, German shepherd dogs and golden retrievers) and entropion in bulldogs. However, it should be noted that these calculations were performed only for the few breeds and diseases where UK prevalence data were available. For most inherited diseases in pedigree dogs, prevalence data were unavailable, unreliable or based on a different population. The need for inherited disorder surveillance at the population level (McGreevy 2007) is clear.

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